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

Kalydeco: Why It Matters To My Overall Health Post-Lung Transplant

By Jen Weber

I started taking Kalydeco about a year ago—just as I had passed the third anniversary of my second bilateral lung transplant. Interest in Kalydeco as a potential addition to my treatment plan began during my routine lung transplant appointment, when I began asking whether these newer CF medications could be of benefit to me. My transplant team encouraged me to pursue evaluation with the adult CF Center, where I was not currently continuing to be monitored since my first transplant in 2010.

I was able to return to my local adult CF clinic, which had established clinic hours for post-transplant CF patients, and learn about the potential CF therapies that might be worth pursuing. In my case, Kalydeco was one treatment option. The chief objectives of beginning Kalydeco were to improve my continued sinus symptoms—thought to be contributing to bouts of lung infections, rejection and inflammation episodes, and to hopefully improve my weight.

I am a CF adult who has had several sinus surgeries throughout life and post-transplant occasionally continued to need IV antibiotics to combat pseudomonas infections. I also have chronic pancreatitis, and anything that might help clear out the sludge in my pancreatic ducts was thought to be beneficial. I did happen to locate two isolated studies of CF patients specifically observed for sinus improvement while on Kalydeco (all thirteen patients were pre-transplant status with a G551D mutation). In these studies, they did appear to improve, which was an encouraging support of my goals.

JEN WEBER

With these objectives in mind my transplant team of doctors, with assistance from the CF clinic pharmacist, first looked at my extensive post-transplant medication list to determine if adjustments would be necessary. I would need to reduce two of my immunosuppressive agents but otherwise nothing extensive. Then we conducted baseline CT scans of my sinuses, and I had a panel of lab values

Continued on page 10

INSIDE THIS ISSUE

CF ROUNDTABLE
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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

Glory be! I hope winter will be over soon. I am so tired of the east wind roaring down the Columbia River gorge. When there are temperatures in the 20s or below, it gets cold. I know it has been far colder than that in many regions of this country this month. I don't like cold, especially when it is accompanied by snow. Yikes!

Come on spring.

Now on to the news of this issue of *CF Roundtable.* The first piece of news is sad. **Anne Williman**, who was a Director of USACFA, died. Her obituary is on page 37. She was a fine woman and we all miss her.

I hope you have read **Jen Weber's** article on the cover. She has interesting information on the use of Kalydeco after lung transplantation. She asked one of her physicians, **Dr. Cynthia Brown**, to write of her role in Jen's use of Kalydeco for our newsletter. Dr. Brown kindly did that and you may check it out on page 12.

Beth Sufian uses "Ask The Attorney" to explain the importance of keeping accurate medical records and also answers questions about Social Security disability coverage and reviews.

The Focus topic is: Using Non-traditional Medicine Or Treatments. **Amy Braid** tells of using cranberry products to combat urinary tract infections. **Andrea Eisenman** discusses Reiki treatments and what a difference they have made for her. **Linda Stratton** relates her experiences with the use of natural medicine. I talk of using kindness as therapy, as well as a couple of other alternate therapies, in "Speeding Past 50."

You'll find an interesting interview of **Briauna Peters** done by **Aimee Lecointre** in "Active For Life." **Dana Giacci** uses "Parenting" to talk of the power of playing and using play with your children as therapy.

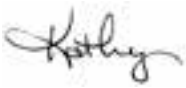
In "Voices From The Roundtable," **Devon Wakefield** writes of his move from the San Francisco Bay area to Seattle, Washington. He offers some tips to help make such a move easier. **Jeanie Hanley** offers some excellent advice about being your own advocate or having another person act as an advocate for you.

Once again, **Laura Tillman** has compiled a long list of "Information From The Internet," where you will find news of clinical trials and new research. In "Searching For The Cure," **Reid D'Amico** writes of the importance of all the research that is being done on aspects of CF other than CFTR.

Julie Desch, in "Wellness," tells us of her invention of "medishaking" or meditating while doing the Vest. **Isabel Stenzel Byrnes** uses "Spirit Medicine" to talk of willpower and its importance in our health.

As we start another new year (our 27th!), let us all remember to be kind to one another and to do what we can to make life easier for all of us.

Please stay healthy and happy.



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Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Viral vector-based approaches could improve effectiveness of CFTR gene therapy

Cystic fibrosis is characterized by accumulation of thick mucus in the lungs and is associated with a high incidence of bacterial infection. Mutations in the gene encoding CF transmembrane conductance regulator (CFTR) underlie the disease. Gene therapy to deliver a normal copy of the CFTR gene has shown promise in both pre-clinical models and clinical trials; however, current gene delivery methods are inefficient and do not result in sustained expression of functional CFTR in the airways.

Two studies report the development and use of viral vector-based delivery of CFTR in pig models of CF. A team developed a lentiviral-based vector that was delivered through the nose to newborn CF pigs. Analysis of these animals revealed evidence of functional CFTR expression in the airways. A second team generated an adeno-associated virus (AAV) that homes in on the pig airway to mediate expression of functional CFTR in the airways of one-week-old CF pigs. Together, these reports indicate that viral vector-based approaches could potentially improve the effectiveness of CFTR gene therapy.
http://tinyurl.com/j4z9eyq
AND
http://tinyurl.com/hwldett

Exemplar Genetics ExeGen® Cystic Fibrosis Research Model Utilized in Development of New Gene Therapy Treatments

Exemplar Genetics announced two publications demonstrating the potential of ExeGen® CFTR miniswine research models to help define and develop effective gene therapies for cystic fibrosis. In the first publication researchers utilizing Exemplar's research models demonstrated that delivery of gene therapy showed an increase in bacterial killing as well as partial physiological improvement in cystic fibrosis (CF). This proof-of-concept study provided the first evidence of CFTR correction by a lentiviral vector in a large-animal model.

The second publication describes an adeno-associated virus (AAV) gene therapy that revealed evidence of functional CFTR expression in the airways of the ExeGen® CFTR models. Use of Exemplar's engineered miniswine mod-

Continued on page 13

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**

Winter (current) 2017: Using Non-traditional Medicine Or Treatments.

Spring (May) 2017: Traveling With CF. (Submissions due March 15, 2017.) What experiences have you had traveling with CF? Are there any pitfalls that we can avoid? Do you have any tips that you can share? Please tell us of your experiences.

Summer (August) 2017: Problem Solving With CF. (Submissions due June 15, 2017.) Do you have hints for solving problems that arise with CF?

Autumn (November) 2017: Dating And Relationships With CF. (Submissions due September 15, 2017.)

ASK THE ATTORNEY

Medical Records And Increase In Continuing Disability Reviews

By Beth Sufian, J.D.

In this month's column we provide information related to medical records and their importance to a person with CF as well as information related to the increase in Continuing Disability Reviews being conducted by the Social Security Administration (SSA).

Nothing in this column is meant to be legal advice about your specific situation and is meant only as information.

I. Medical Records Are Crucial

Detailed and accurate medical records can help with a variety of issues. Medical records are important for those applying for Social Security benefits or undergoing an SSA Continuing Disability Review. Typically, the basis for an approval or denial of Social Security benefits is found in a person's medical records. In addition, insurance companies are increasingly requesting medical records to determine if coverage for expensive treatment will be provided. Schools and employers may also require medical records if a person requests an accommodation at school or work.

While there is no exhaustive list of what should be included in a person's medical records, it is helpful if medical records contain information about daily limitations that interfere with or prevent a person from working full time or attending school.

Medical records often do not contain details needed to obtain insurance coverage, Social Security benefits or employment or school modifications. It is important to make sure a CF Center records medical issues and limitations on daily life activities in the medical records.

Information such as the time it takes to perform each breathing treatment done in the morning and each treatment done at night; the number and duration of airway clearance treatments; the need for naps during the day; issues with coughing up blood or digestive issues; issues with memory, concentration or stamina, coughing at night or insomnia caused by medication side effects are all important. The time spent managing CF-related diabetes is also important. If diabetes is difficult to control, then it is important that the medical records discuss the problems CFRD is causing for the person with CF.

Any information that supports a finding that the person cannot work full time is helpful for a Social Security application or a Continuing Disability Review.

It is a good idea to keep a log of issues a person is having in between CF Center appointments. A spiral notebook will do the trick. Dates health issues were experienced are crucial. For example: "December 12, 2016 - Hemoptysis. Coughed up a teaspoon of blood." Another example would be: "December 15, 2016 - severe stomach problems requiring rest for two hours mid-day."

BETH SUFIAN

Often a person might forget issues they were having a month before their clinic visit. Sometimes there is so much to discuss at a clinic visit that some things are not discussed and so are not recorded in the medical records. A notebook containing a log of issues a person had will be helpful at the clinic visit. The person can tell the CF Center team member what happened and ask that the information be included in his or her medical record.

A letter written by a physician at the time a person applies for SSA benefits is not as effective as detailed medical records for a 12-month period. Records that document medical issues and issues with performing daily life activities are extremely helpful.

There is no guarantee that including such information in a person's medical records will result in an approval for SSA benefits, insurance coverage for a needed treatment or modifications at school or work. However, it typically helps to have detailed information about a person's medical condition in his or her medical records.

II. Increase in Continuing Disability Reviews

Social Security has had an increase in funding from Congress for the specific purpose of conducting 400,000 additional Continuing Disability Reviews (CDR). Many CF Centers

have reported that large numbers of their patients who receive SSA benefits have received CDRs from SSA in the past three months.

Often, people with CF or their parents do not realize the significance of a CDR and mistakenly believe that completing the CDR paperwork is an easy task with little effect on the ability to maintain SSA benefits. This is an incorrect understanding of the significance of the CDR paperwork. The CDR paperwork is very important.

One answer on CDR paperwork indicating a person is actively playing sports at school or working part time at a physically demanding job can lead SSA to determine that benefits should be stopped.

While answers should be true and correct, it is important to explain if a person with CF is participating in sports but takes frequent breaks or must come home and sleep for two hours after engaging in sports at school. The same holds true for engaging in part-time work. If a person works four hours as a cashier but then goes home and sleeps for two hours and must rest the next day, it is helpful that such limitations are noted on a CDR.

A person can appeal a benefit termination but it typically takes one to two years until a hearing is scheduled before a judge to determine if the termination was correct.

III. SSA Rules Used to Determine Eligibility

ALL cases pending at SSA after October 7, 2016, will be determined using the new CF medical eligibility rules. When new medical criteria issued by SSA specifically state that all pending cases will be decided under new rules, that means that all currently pending cases will be decided under the new rules.

Some people are confused and assume that the new rules apply only to new applications for SSA benefits. This assumption is incorrect. In the past, the SSA general rule has been that a Continuing Disability Review is decided based on the rules in effect when the initial application for benefits was filed.

HOWEVER, if the new SSA medical criteria specifically states that all pending cases will be decided under the new rules, then CDRs are determined using the new rules.

All of the CDRs handled by the CF Social Security Project® are being determined using the new medical criteria. These cases are spread out over 26 states. All local SSA offices have informed the CF Social Security Project that, "of course we are using the new rules to determine eligibility in a CDR for a person with CF because that is what the new medical criteria tells us to do." Every SSA Regional Chief Judge has confirmed that SSA offices are using the new CF medical criteria to determine eligibility under a CDR.

It is very important that people with CF and their CF Centers understand which medical criteria will be applied to their CDR. If a person with CF thinks that the old rules will be used to determine eligibility during a CDR, then the person will not be providing SSA with the information needed to determine whether benefits should be continued. If a person does not meet the new rules, the person must show his condition is medically equivalent to the new rules.

The CF Social Security Project will continue to represent individuals with CF in an initial application or in a Social Security Continuing Disability Review at no cost to the individual. While funding for the CF Social Security Project comes from the CF Foundation, the CF Social Security Project is operated by Sufian & Passamano, a law firm in Houston, Texas. The attorneys at Sufian & Passamano, LLP, are not employees of CFF.

The CF Legal Information Hotline® continues to provide information about Social Security benefits, insurance, employment and education to the CF community. The Hotline is sponsored by a grant from the CF Foundation and can be reached directly at 1-800-622-0385 or by e-mail at: CFLegal@suffianpassamano.com.

A person can contact the CF Legal Information Hotline directly and there is no need for an authorization form. The CF Hotline can speak to anyone. A parent can call for information related to his or her adult child. No release is needed to speak to the CF Hotline.

The CF Hotline received over 1,000 calls in the four weeks post-election. This is the largest number of calls the CF Hotline has received in a four-week period in its 18-year history. It is best to e-mail the CF Hotline and set up a specific time to speak to an attorney.

People are concerned the Affordable Care Act (ACA)will be repealed and that subsidy help with premium payments will be discontinued soon. People are also concerned that the Medicaid expansion allowed under the ACA will be stopped, leaving many adults who have low income and assets without access to Medicaid benefits. The CF Hotline can provide information related to strategies people with CF used in the past to obtain or maintain insurance coverage before the ACA was in effect.

The CF Social Security Project handles cases for people with CF from around the country and so is able to assess trends quickly. Watch the *CF Roundtable* blog in the coming months for updates on how SSA is interpreting the new rules. ▲

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

SPIRIT MEDICINE

The Spirit Of Willpower

By Isabel Stenzel Byrnes

"Mental health is based on a certain degree of tension: tension between what one has already achieved and what one still ought to accomplish." Viktor Frankl

Hi there! I'm back! I missed writing "Spirit Medicine" for the Autumn issue of *CF Roundtable* because I was preoccupied with a licensing exam that I took just around the time of the deadline for articles. Thankfully, I passed! For about 18 years, I've wanted to obtain my license in clinical social work (LCSW). For many years, the LCSW was a distant hope and dream, because cystic fibrosis (CF) caused many starts and stops in my career. So for this article, I'd like to write about the impact of goal setting and willpower on our spirits. Hopefully, you'll get this issue in the early part of 2017, and this topic will be relevant to you for your New Year's resolutions!

So what's up with having a goal? A goal is something to look forward to, to exert effort toward and to sustain hope. For many years, I had simple health goals...Staying out of the hospital. Gaining a pound. Taking a walk. After my transplant, my goal was to walk a mile in three months and run a mile in a year. It feels good to fulfill a goal. It means I have agency, or the ability to have control over something.

I've had a bucket list since I was a very young child. The term "bucket list" was popularized by the 2007 film with Jack Nicholson and Morgan Freeman; two men with terminal cancer travel the world fulfilling their goals. My bucket list has included big things like graduating from college; getting married; having a career; running a mile; surviving to be 30, then 40, now 50; playing the bagpipes; going to Machu Picchu, and the list goes on and on. Most of these goals I've managed to do and I know I'm very, very fortunate. I feel a sense of completion to cross something off my "bucket list" and move on to something else. It lifts my spirits and makes me feel that I'm fully engaged in life.

Each time an item was crossed off the list, I felt compelled to add a new goal onto the list. When my sister went into rejection after her first transplant, she told me, "I have accomplished all my goals. I'm done." Eventually, she chose to have a second transplant and survived seven more years. But since then, I have feared that if we have no more goals, our life energy will dissipate, and we will die. Goal setting lights an inner fire to keep going.

ISABEL STENZEL BYRNES

As I get older, like the opening Frankl quote above, my mental health depends on reviewing the goals I have already accomplished and the new ones I'd like to add to my list. I remember my sister Ana's last coherent words before she died: "I've lived a good life. I've done everything I've wanted. I even got married." Goal setting and goal fulfilling can help with acceptance at the end of life. It can fill the end with gratitude more than regret. Developmental psychologist Erik Erikson called this "ego integrity" instead of "despair." Death is more tragic when people with CF die right in the middle of college or who leave behind a fiancé. Life seems incomplete.

Now for those of us who are in the midst of living with CF, let's explore how it feels to not fulfill a goal. It is hard. This feels like failure, or a test of patience, or an injustice for things getting in the way. When external issues impede our goals, we have to accept that many things are not in our control. But one thing that is in our control is willpower.

I'd like to explore the issue of willpower. Willpower is defined as the ability to control our impulses for immediate comfort or pleasure for long-term goals. I'm going to be honest here. My impulse, every day, is to stay in bed. I want to lie on the couch, to stare at the wall. I deal with fatigue every day, despite what I post on Facebook. But I know I have to get up, take my meds, deal with blood sugar, exercise, do my aerosols and sinus flushes and keep going.

One of my inspirations is Kelly McGonigal, a Stanford educator, who

wrote a book called *The Willpower Instinct.* She states that the human mind is made up of three opposing forces: the "I will" mind, the "I want" mind and the "I won't" mind. Like the general population, people with CF vary in their degrees of willpower. Some do their treatments gladly; others need a push. Some don't exercise; others run marathons. Willpower is harder to muster the crummier you feel.

> *"When external issues impede our goals, we have to accept that many things are not in our control. But one thing that is in our control is willpower."*

McGonigal says willpower is influenced by prior experience withholding or denying gratification. I'm going to extrapolate and say people with CF develop willpower muscle over their lifetimes. Our effort to do treatments, to put health first, to exercise and maintain weight, to manage our time and priorities, molds our willpower. So what control do we have over our willpower? What can our spirits do to cultivate willpower?

McGonigal has studied problem behaviors like smoking, drinking, procrastination, gambling, overeating etc. She is an academic, so she has conducted or reviewed numerous research studies looking at cravings, self-talk, delayed gratification... all the things that contribute to willpower. She found that people with the most willpower have certain common traits:

1. They get plenty of **sleep**. People who get a good night's sleep have a bigger and denser prefrontal cortex: the part of the brain that focuses on long-term goals and core values that aid in willpower. Even power naps help to recharge willpower.

2. They eat a **balanced diet** with low glycemic foods. Spikes in blood sugar prevent the brain from being able to focus. Having regular blood sugars sustains energy and helps the brain pay attention.

3. They **exercise** regularly, which increases circulation and helps to focus the prefrontal cortex.

4. They **meditate** regularly: this cultivates awareness and builds more gray matter in the prefrontal cortex. Meditation reduces cortisol and helps with emotional control. Meditation enhances memory, attention and the ability to ignore distractions.

5. They make friends with their **future selves**. They imagine themselves in the future and what they would be like if they did or didn't practice the desired behavior now. This motivated people to take better care of themselves now. An exercise she used for this study was to have participants write a letter to themselves from their future self to current self. Those who were more similar in the present to their imagined future self had greater willpower.

6. They get a lot of practice with **decision-making** and consequences of willpower. In other words, prior successes with willpower lead to greater successes in future willpower challenges.

7. They predict **failure** and practice "defensive pessimism," which is the anticipation of failure but taking the steps to prevent it. Imagining failure helps motivate people better than imagining success.

8. They "**surf the urge,**" which means when their willpower is challenged, they practice mindfulness, simply noticing the discomfort, acknowledging the feelings that come up and they are able to tolerate those feelings and not act on those feelings or impulses.

9. They practice **self-compassion**. Self-criticism, guilt, regret and feeling bad about yourself undermines willpower. Her studies show that these behaviors stress you out and make you more likely to repeat a negative behavior. In other words, being kind to yourself and forgiving yourself when failures happen will help your willpower. Give yourself a compassionate message; she suggests you talk to yourself like you'd talk to a friend. She also encourages you to imagine "common humanity"— which means embracing that all people struggle with and cope with what you are going through.

10. They have good **distress tolerance** or the ability to stay put when uncomfortable. She asked people to hold their breath for as long as possible and those who could tolerate discomfort longer also tested higher for willpower in other tasks.

11. They have **new goals and new challenges** constantly as a great way to boost their willpower. They regularly move out of their comfort zones into personal and creative challenge. Being in a deep creative state, called a "flow state," helps to grow willpower. (Like how I get writing these articles! Shameless promotion: write for *CF Roundtable!*)

Okay, do these traits look familiar? To survive with CF, we need these willpower traits! I think the vast majority of adult readers could relate to having many if not all of these traits. Imagining yourself as a sick older adult might motivate you to do treatments now. Tolerating discomfort is something many of us are pros at! McGonigal says many of these habits condition the brain so they become automatic and

Continued on page 29

SPEEDING PAST 50
Kindness As Therapy

By Kathy Russell

Wow! This winter has started out with a bang! Talk about cold. I am so happy that we have a wood stove that keeps our house toasty warm. When that cold wind blows out of the Historic Columbia River Gorge, the warmth can be sucked right out of our house, but the wood stove keeps us warm.

I always have been a pretty "by the book" kind of person, when it comes to my healthcare. I am not apt to try new things or things that are not usual care. However, on occasion, I have tried a treatment that was different or new.

I had severe sinus disease for the first five decades of my life. When I was 51, I had my last sinus surgery. It was my third sinus surgery in not too many years. The first were the old-fashioned Caldwell-Luc procedure. The final surgery was done with a laser. The laser seemed to do a better job of cleaning everything out.

My doctor suggested that I irrigate my sinuses with a tobramycin mixture to help prevent infection. Before I was finished with those irrigations, I asked my doctor if maybe irrigating with a saline and acetic acid solution might help to prevent future sinus infections. Since we use acetic acid (vinegar) to clean some of our equipment, it seemed to me that it might help to keep our membranes clean.

The doctor was open to the idea and he did some investigating into the use of acetic acid in solution for irrigation. He discovered that there were drops for use in the nose that were a solution of 2% sterile glacial acetic acid (GAA) in sterile water. Since I wanted to use enough to irrigate my sinuses, little two or three ml vials of the solution weren't going to work. So he ordered GAA and sterile normal saline (NS) to use for such irrigations. He warned me that the solution could be quite caustic and that I should be very careful about my use of it.

I started by using a minimal amount of GAA in NS and then began to increase the ratio of GAA to NS. I continued to increase the amount of GAA until I reached my level of discomfort. Then I backed off a little on the GAA. I have continued to irrigate with that solution for the subsequent 21 years. I have had only a couple of sinus infections in all those years. I attribute that to my use of that solution.

I just remembered another way that I use an alternate therapy. When I have pain, I use laughter or contemplation, some might call it meditation, to ease the discomfort. The form of contemplation that I use is quiet music in the background, while I close my eyes and relax. I find that most of my angst eases as the music washes over me and I feel at peace. I am able to let my mind take me away from whatever is bothering me and just let time pass.

Laughter is a great way to ease my pain. I remember a time when I was in the hospital. I had recently had half of my colon removed. I was experiencing some severe abdominal pain. When I called for a nurse, I discovered that they all were busy with a code blue. No one could break away to get a pain med. So I called my husband on the phone. I asked him to make me laugh. I explained to him that I was having pain and no one could assist me right then.

> *"When I have pain, I use laughter or contemplation, some might call it meditation, to ease the discomfort."*

KATHY RUSSELL

He reminded me of a time when our dogs were racing around the house, chasing each other. They would go one way at top speed, until they reached the end of the house. Then they would turn around and the chaser became the chased. They kept it up until they both were worn out. It was a hoot to watch and was funny to remember. As he reminded me of that day, I started laughing and my pain eased. By the time the nurses were free to help me, my pain was so much better that I didn't need any medicine. Laughter really had taken care of the pain.

When I was young, rather than doing chest physio therapy (CPT), I did acrobatics (gymnastics) and took ballet and modern dance classes. I also played

on a volleyball team. These forms of exercise helped me keep my airways clear.

As I got older, I didn't have time for those activities. My normal activity at work seemed to provide me adequate airway clearance, most of the time. When I needed more clearance, I had someone do conventional CPT. Since I worked in the hospital that treated all the young people with CF in my state, I could just stop in the PT department on my way to work and get my CPT. It really helped to be able to access the therapy when I needed it. It was one of those professional courtesies that can be so nice. Once I no longer had that available, I learned how to clear my airways with breathing exercises. I still use those, even though I do medicines in nebulizers.

I never have tried acupuncture or chiropractic medicine, although I know some people who have had very positive results from either or both therapies. I think we must be open to trying new things and using new practices. Some treatments that once were considered "alternative" therapies now are considered usual practice. Conversely, some practices that were absolute when I was young are no longer done. Thank heavens mist tents no longer are used. Also, our plastic nebulizers are so much safer to use than the old glass nebulizers we used to have.

Caregivers now are accustomed to fitting the treatments to the patients, rather than making patients do a certain therapy because some guideline says that is how it must be done. Let's hope that trend continues.

As we age, we learn new things about ourselves and our CF. I have found that I no longer am able to discern when I am getting sick enough to need intervention by my docs. Paul, my husband, has to almost get angry with me as I am getting ill, because I just don't think I am that ill. I tend to wait longer than I should to call my doctor. This is not good and I have to work on doing that better.

During the last six months of 2016, I fought with my lung bugs and they finally knocked me down. I had a couple of bouts of large hemoptysis that caused me some angst. Even with bronchoscopy, the docs were unable to see where the blood was originating. At least it stopped, after a few days. I turned my O_2 up to 4L/minute and my sats still were dropping low. One day, when I was feeling a little off, I checked my saturation and it was down to 73! I couldn't ignore that.

After another hospitalization and three weeks of home IVs, I am back to 3L/minute and doing okay. My sats are staying in the 90s and my heart rate is much better than it had been. I don't know how many more times I will be able to dodge a bullet, but I am going to keep on trying.

I will continue to take good naps in the afternoon and get my eight hours of sleep at night. There is nothing I have to do that is so important that will keep me from getting enough rest. When I am well-rested, I enjoy life and am able to do a few things that bring great pleasure.

Since none of us know how long our lives will be, I suggest that we put a little gusto into every single day. Do something that you enjoy, even if for only a few minutes. You should have a little pleasure in every day. Be sure to let those who are important to you know that you care about them and appreciate them. This is especially true of our spouses. We tend to start taking them for granted and forget to thank them and compliment them on jobs well done.

Although none of us can change the world situation, each of us can make our little corner of the world a kinder, more peaceful spot. Practice kindness, it is a great form of therapy. You will be amazed at how much better you will feel, when you are trying to be kind.

Until next time, stay healthy and happy. ▲

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

Cystic Fibrosis Mothers

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

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

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

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

WEBER continued from page 1

both to see what my current pancreatic enzyme levels were and ensure none of the common side effects of Kalydeco would be a concern (chiefly my liver functions).

After discussions with both the transplant team and CF clinic doctors and a thorough medical exam, I was ready to begin Kalydeco. I was surprised and relieved to find that even as a post-transplant CF patient, all my private insurance required for approval was documentation of my CF diagnosis with the genotype (for me, DF508/R117H) indicating its appropriateness.

Vertex offers extensive patient support for individuals taking Kalydeco, and the next step was a phone call with their counselor explaining how to take it and assisting with my insurance to find the correct distribution pharmacy. My dosage of Kalydeco was started at the standard adult dosage, 150 mg every 12 hours with a high-fat containing snack. This was easy to incorporate into my 12-hour dosing schedule with my immunosuppressive medications. Kalydeco also places restrictions on eating food containing grapefruit or Seville oranges, which is similar to my transplant meds that require that caution as well.

> "On a daily basis, the reduction in chronic sinus drainage has been the greatest sustained benefit of Kalydeco for me."

My initial observations were essentially nothing—which I took as a relief! My lungs felt OK, no stomach upset, headache or dizziness. After taking Kalydeco for about two months, I was encouraged that I had gained approximately seven pounds with little effort. Within another month—about month three—I did develop a severe acute pancreatitis attack requiring a lengthy hospitalization over several months. I briefly stopped the medication to see if it might be contributing to my symptoms, but it did not appear to be a factor and I re-started it.

At the six-month mark, my sinuses were re-evaluated—chiefly because I was

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

Michelle Allen
Portland, OR
65 on September 14, 2016

Johanna Libbert
Richland, IN
50 on June 20, 2016

Debra Radler
Roselle, IL
54 on May 31, 2016

Wedding

Michelle & Gary Allen
Portland, OR
20 years on April 27, 2016

Susie Baldwin & Adam Levy
Los Angeles, CA
20 years on October 13, 2016

Victor & Linda Roggli
Durham, NC
25 years on April 27, 2016

Transplant

Cris Dopher, 45
Brooklyn, NY
Bilateral lungs
3 years on December 13, 2016

April Hansen, 23
Brooklyn, NY
Bilateral lungs
3 years on December 13, 2016

Joy Heinsohn Smith, 41
Boca Raton, FL
Bilateral lungs
1 year on November 10, 2016

Johanna Libbert, 50
Richland, IN
Bilateral lungs
2 years on October 20, 2016

experiencing pulmonary issues (not sinus). It appeared some of the sinus cavities were clearer, but I did still have issues with recurrent infection causing inflammation. I was also continuing to have occasional sinus headaches, but the daily sinus drainage I used to manage was somewhat improved. I agreed to another sinus surgery at that point and since that time my recurrent headaches are much reduced.

As I review the year, I believe Kalydeco was a sensible choice for me. I have only been hospitalized for one lung infection and acute rejection episode and my lung functions finally stabilized—I have required antibiotics for recurrent lung infections a few times as an outpatient, but I believe these episodes were relatively minor. I cannot attribute all my lung stability with any certainty to Kalydeco, but I do believe the reduction in sinus infections and drainage is contributing to that significantly. On a daily basis, the reduction in chronic sinus drainage has been the greatest sustained benefit of Kalydeco for me—my initial weight gain seemed to level off and I have now lost most of it. My liver labs are stable, and I've experienced none of the common side effects of Kalydeco.

I believe when sinuses are healthy, lungs are healthy—and that seems to be the case, with me. I will keep an eye on the continued pipeline of medications being developed and researched for individuals diagnosed with CF—whether we are pre- or post-lung transplant. There are so many factors that contribute to overall good health.

While it is logical to focus on the pulmonary issues so often related to CF, I was glad my team of doctors were willing to look at all the things I can do to support my healthier lungs by having a healthier body systemically. Coordination with my CF team and transplant team was essential not only for beginning my treatment with Kalydeco, but I believe continuing to ensure I benefit from hearing and receiving all healthcare perspectives necessary to collaborate in my care. For now, continuing to include Kalydeco in my cocktail of daily medications seems to be working for me. ▲

Jen is 43 and has CF. She is a Director of USACFA and is the Secretary. Her contact information is on page 2.

Mailbox

Brian Marceau is an inspiration to all of his family. Thanks for all I have learned from *CF Roundtable*.

Cathleen Marceau
Coeur d'Alene, ID

Debbie Radler (my daughter) is very strong, very positive and never, ever, gives up. I am so proud of her.

Loretta Mulatz
Bloomingdale, IL

Keep doing your thing.

Mary Lou Figley
Monument, CO

Keep up the good work; I love this organization!

Lisa Cissell
Bardstown, KY

I am so excited to reach age 65 and still be productive.

Michelle Allen
Portland, OR

I have a 13-year-old with CF, but still find *CF Roundtable* very helpful and informative. Thank you!

Ann Black
Royersford, PA

Keep up the great work! This is a terrific publication.

Sara Kominsky
Blacksburg, VA

Thank you.

Susie Baldwin, M.D., M.P.H.
Los Angeles, CA

Keep up the inspiring work! ☺

Joan Finnegan Brooks
Osterville, MA

My husband always loved *CF Roundtable*. Our two sons who have had bilateral lung transplants are doing well.

Dorothy Stewart
Hobbs, NM

Thank you for all you do.

Ted & Susan Tickell
Bridgewater, CT

CFTR-Modulating Drugs After Organ Transplantation

By Cynthia Brown, M.D.
Associate Professor of Clinical Medicine
Director, Adult CF Program
Indiana University

On the cover of this issue, Jennifer Weber writes about her experience using ivacaftor (Kalydeco®) after lung transplantation. I have the privilege of being part of Jen's care team at Indiana University along with my transplant colleagues. In November 2015, Jennifer returned to the Adult CF Center. She had not been seen by anyone in the CF Center for many years following her transplants, and her care was guided by the transplant team. Jen returned to clinic with a very specific question: Would she benefit from ivacaftor following her lung transplantation? Her essay describes her experiences with the medication with minor improvements in her sinus disease and no significant improvements in her recurrent episodes of pancreatitis. As a CF care provider, I asked to write a companion article to further detail issues regarding CFTR-modulating drugs after organ transplantation, and I will discuss both ivacaftor (Kalydeco®) and lumacaftor-ivacaftor (Orkambi®).

1. There is no published medical literature that supports use of these drugs following transplantation of any organ.

When Jen proposed using ivacaftor last November, I did not say yes immediately. I first attempted to review the medical literature (which was a quick study, given that nothing had been published on use after transplant). I then proposed the question on a CFF-sponsored listserv and received little response. In fact, it was Jen herself who had seen a Facebook post on a patient transplanted at Columbia University in New York who had continued to use it. So I contacted the physician there who reported that it was being used to assess response on acute pancreatitis. Then I reviewed the medical literature for effects of ivacaftor on other symptoms after transplant. My clinical experience led me to believe that ivacaftor would be beneficial for chronic sinus symptoms and, indeed, a few articles support this although there are no articles about pancreatitis and ivacaftor. There is even less information about the effects of the combination drug lumacaftor-ivacaftor (Orkambi®) on non-pulmonary manifestations of CF, as this drug has been FDA-approved only for about 18 months. Data published to date have focused on pulmonary effects, exacerbation rates and weight changes. I do want to point out that there is no role for using the CFTR-modulators for pulmonary effects after lung transplantation. The medications would be expected to have no effect since the transplanted donor lungs do not have the same defects in CFTR function. Finally, I could perceive a role for either CFTR-modulator for pulmonary function after transplant of other organs, including liver and pancreas, but there is nothing published about this either at this time.

So ultimately, Jen, her transplant team and I decided to give ivacaftor a trial based primarily on what we know

Drug	Ivacaftor (Kalydeco®)	Lumacaftor-Ivacaftor (Orkambi®)
Azole antibiotics (Fluconazole, Itraconazole, Voriconazole)	↑ Ivacaftor level, dose reduction required	↑ Ivacaftor level, dose reduction required
Macrolide antibiotics (clarithromycin, erythromycin, but NOT azithromycin)	↑ Ivacaftor level, dose reduction required	↑ Ivacaftor level, dose reduction required
Narcotic analgesics (morphine, hydrocodone, etc.)	↑ exposure to narcotics, may require dose adjustment	↑ exposure to narcotics, may require dose adjustment
Sirolimus	↑ sirolimus levels	↓ sirolimus levels
Tacrolimus	↑ tacrolimus levels	↓ tacrolimus levels

> My clinical experience led me to believe that ivacaftor would be beneficial for chronic sinus symptoms.

about its mechanism of action and potential effects on sinus disease and chronic pancreatitis. However, we would do this only if we could safely manage drug interactions with her needed transplant drugs.

2. Drug interactions between CFTR-modulators and many medications used in transplant are common.

Once we decided that ivacaftor may be beneficial clinically, the next step was to determine if it could be used safely. Jen's most recent transplant was performed in 2012. So at the time of discussion, she was at a point where there were not many anticipated changes in her routine post-transplant medications. The consideration for use of these medications is potentially more problematic early in the post-transplant course when there are more potential drug interactions. Moreover, lumacaftor-ivacaftor (Orkambi®) has even more drug interactions than ivacaftor due to the way it is metabolized in the liver. I have included a table that lists the interactions between ivacaftor, ivacaftor-lumacaftor and commonly used medications after transplantation.

3. A CF care team can be useful partners following lung transplantation.

I came to Indiana University in July 2014 as the Adult CF Director. Prior to my arrival, CF patients who had a lung transplant typically did not return to the CF Center for any further care. In many lung transplant centers, this seems to be the typical practice pattern and some transplant centers have physicians on the transplant team who are also members of the CF care team. However, many lung transplant physicians do not have significant CF care experience. Since arriving at IU, we have established quarterly clinics for the care of CF patients who have had a lung transplant. Typically, I see transplanted CF patients only once or twice yearly. We schedule these patients on days when no other CF patients will be in clinic, for infection control purposes and to limit any potential exposure to infectious organisms. It is important to consider that while the lungs are healthy, other organs continue to have CF-related difficulties. Specific issues that I frequently find in the post-transplant CF patients are related to the GI tract and sinuses. My transplant colleagues are aggressive in treating sinus disease, but having the multidisciplinary CF team including a CF-trained dietitian can be very important for addressing GI issues. In addition, the CF team is best prepared to answer specific questions about new medications, such as the one proposed by Jen. Finally, an added incentive for me is that it brings the now-healthy lung-transplant patient back into our clinical area so that the entire CF team can see the life-changing efforts of the care we provide.

In summary, there may be a place for CFTR-modulators in the care of the post-transplant patient. However, the patients that may best benefit are not yet known. It is very important to coordinate care between the CF team and the lung transplant team prior to consideration to determine potential benefits, risks and drug interactions. ▲

Dr. Brown may be contacted at: cyndbrow@iu.edu

TILLMAN continued from page 3

els offers a platform with greater anatomical, physiological and genetic similarity to humans, mitigating the lack of translation with murine systems due to differences in size and metabolism.

In addition to its utilization for advancing gene therapy, the ExeGen® CFTR model has also been employed to investigate small molecules in the treatment of CF.

http://tinyurl.com/z6dl33r

Cystic Fibrosis Inflammation Eased by Arthritis Treatment in Study

Abnormal activation of inflammasomes, complexes that lead to the production of pro-inflammatory molecules in response to a pathogen, often contributes to respiratory infections and pathologic airway inflammation. In a recent study, Italian researchers looked at how two distinct inflammasomes, NLRC4 and NLRP3, participate in cystic fibrosis (CF)-associated lung infections, showing that NLRP3 contributes more actively to harmful inflammatory responses. The study also found that NLRP3 activity can be impaired by Kineret (anakinra), an approved rheumatoid arthritis treatment, revealing a promising therapy approach for CF patients.

To understand the role of NLRP3 and NLRC4 inflammasomes in CF, the team infected CF mice with Aspergillus fumigatus and Pseudomonas aeruginosa. Results revealed that both inflammasomes are involved in pathogen clearance, but NLRP3 was more commonly associated with harmful inflammatory responses in CF than NLRC4. This NLRP3-dependent response was particularly evident when the NLRC4 response was absent, suggesting that the

Continued on page 15

FOCUS TOPIC

USING NON-TRADITIONAL MEDICINE OR TREATMENTS

Keeping It In The Family

By Linda Stratton

When my sister, Cheryl, graduated from Bastyr University with her doctorate in Natural Medicine in the spring of 2001, we were both overjoyed. My sister, because after five long years of hard work, she was following her passion of healing and wellness of the human body through natural means. Me, because I hoped her gained knowledge would be a significant weapon in my fight against cystic fibrosis (CF).

In the beginning, after much research, we decided the only way it could work to fight my CF was if we used a combination of traditional medicine with natural medicine support. We started out slow, but there were and still are many ways in which natural medicine has helped me on this roller-coaster ride of a life with CF. We've been looking for a strong enough, natural-based antibiotic to get rid of pseudomonas, but haven't quite been successful.

The first thing she did was get me on probiotics, considering all the antibiotics I was taking. I prefer Culturelle—the change in my digestion and bowels was immediate. This was combined with good nutrition. We tried many different scenarios; I must admit I'm not the best patient in this category. I guess I feel like I've given up everything in life that gives me pleasure...I want what I want when it comes to food. I do try to follow most of what she tells me, however, I WILL NOT GIVE UP DARK CHOCOLATE!

She helped most when I was diagnosed with CFRD (cystic fibrosis-related diabetes). We went over the lists of healthy carbohydrates, simple carbs, low-glycemic carbs etc. She also taught me about protein-carbohydrate ratios and how important healthy fats are to one's diet. Her help was invaluable.

For several years now, I've been drinking a recommended protein drink with all of the vitamins and minerals my body needs. Most times my blood work comes out beautiful. Although, you must know and keep in mind—I don't suffer mal-absorption problems in the normal CF way.

For years and years, Cheryl has given me my own personal homeopathic remedy for depression and emotional well-being. This is done through an in-depth interview, with questions about physical, mental and emotional states—then adding several hours of research. What a blessing this has been! I keep track of my doses and can usually feel when one is due. It keeps me from feeling overwhelmed and depressed through my life's difficult times.

Several years ago, I started taking N-Acetylcysteine oral tablets prescribed by Cheryl. This natural medicine was supposed to help thin the mucus in my lungs. I tried it and stopped many times—I had to take several tablets several times a day for it to be effective, which I couldn't seem to accomplish.

DR. CHERYL GROSSHANS AND SISTER, LINDA STRATTON, ON THANKSGIVING DAY.

She used to say, "If we could somehow get it directly into your lungs, it would be effective." Coincidently, my CF doctor prescribed Mucomyst, which turned out to be the acetylcysteine Cheryl was trying to give to me all along. I continue to use it currently and am totally aware it's considered "old-fashioned." Hypertonic saline was never a favorite and the Mucomyst works much better for me and I'm thankful to have it—I guess I'm just an old-fashioned girl. This is a good example of how both types of medicine can coincide and work together.

Another way natural medicine has helped me in an extreme way is during cold and flu season. I use two different products if I have a hint of either. One is a botanical tincture made especially by Pharmaca Integrative Pharmacies called "Lung & Sinus Blaster," which does just that. It truly works to soothe my sinus and lungs, also thinning and helping to remove mucus. I call it my "heading it off at the pass" medicine—catching it before an exacerbation gets started. In addition, I use "Alkalol," a natural soothing mucus solvent and cleanser that's been around since 1896; just one tablespoon in my sinus rinse does the trick, and it's sooooo soothing. You can find Alkalol in your local grocery store, health-food store or a place like Walgreens. I also use various natural teas: Breathe Easy by Yogi Tea, and Deep Lung and Gypsy Cold Care from Traditional Medicinal Teas. You can find them at any natural food store.

Finally, working with Cheryl has given me peace of mind in a way that nothing else could. During an emergency situation, she treated my kitty, Bruno, and probably saved his life. After being out of town on business, I came home to a very sick cat. My pet sitter didn't notice his unusual behavior. I knew right away and, because it was a weekend, called my sister. She came with several homeopathic remedies and worked on him most of the night. He survived, and I will forever be indebted for her knowledge and kindness.

In my experience, I'm thankful to have a CF doctor who has treated me for over ten years and who gives me liberty to experiment with and try alternative medicine. Not once has it interfered or caused a problem with my traditional regimen. She is very gracious and understanding when I tell her what we are up to—I believe she knows it has helped me stay healthy.

Just as in my journey with traditional medicine, I have to pick and choose what I believe is best for me. Cheryl has showered me with many bottles of plant-based medicines over the last 15 years, some have worked and some not so much. I'm willing to try just about anything, yet, if I don't feel a significant change or get the result we are looking for, I stop using it. My CF doctor will testify to that as well. I've come to believe, less is better—at least for me. This applies to both traditional and natural medicine. I'm ultra-sensitive to numerous medicines used to treat CF and can also respond negatively to some of the natural remedies. My doctor and sister both know the frustration in the war with my immune system.

I'm a firm believer in using natural medicine when you can and only bringing in the big guns of traditional medicine when necessary. I will testify, however, that nebulizer treatments are a must if a person wants to stay ahead of the game. I can't technically say my longevity is due to my willingness to take advantage of the good in both traditional and natural medicine…is it just genetics? I feel truly blessed either way! ▲

Linda is 62 and has CF. She lives in Denver, Colorado, and looks forward to the change of seasons with the clear, crisp air it brings. She is retired from many years in the restaurant business. She is single, without children, but enjoys 11 nieces and nephews, and 11 great-nieces and great-nephews. You may contact her at: linstratt@gmail.com

TILLMAN continued from page 13

production of an interleukin-1 receptor (IL1-R) antagonist by the NLRC4 pathway could be impairing NLRP3. Consistently, the researchers found that inhibiting the IL-1βsignaling with the approved IL-1R antagonist Kineret could improve the harmful effects of NLRP3. In fact, CF mouse models treated with Kineret showed significantly increased survival to P. aeruginosa infection and lessened bacterial burden, neutrophil (a type of innate immune cell) recruitment and lung damage.

http://tinyurl.com/zj7v4pj

New research sheds light on "gender gap" in cystic fibrosis

In this current study, a multi-university group explored a crucial protein involved in the transport of ions across biological membranes. The ion transport system investigated in the current research involves a critical protein known as KCNE3, which helps properly regulate ion transport. KCNE3 forms a complex with a partner protein—KCNQ1, which acts as a channel for ions. Together, they form a finely tuned gateway, permitting or blocking ion transport in order to maintain salt and fluid homeostasis.

Among its many functions, KCNE3 regulates the flow of potassium (K+) ions that impact transport of chloride

Continued on page 17

Alternative Therapy For Urinary Tract Infections

By Amy Braid

With the amount of medications we take with CF, sometimes it is great to look into alternative or natural therapies in order to reduce the amount of drugs we ingest. I for one take a lot of supplements—vitamins E and D, multi-vitamins, calcium and cranberry. The cranberry I take for my ridiculous susceptibility to urinary tract infections (UTIs).

Back in March 2015 I had a REALLY bad UTI. I won't go into details, but even the nurse at my PCP office was shocked! I was also on a study for colon cancer at Dana Farber so I had to check in with the study coordinator, who asked me if this was my first UTI. I of course said, "Yes," because I have never had had unexplained blood in my urine before. She decided to check my chart to make sure and found that I had nine other UTIs in three years. That was the first time I had heard about my previous UTIs.

She gave me dates and they all lined up with hospitalizations. And when I thought about it, it all made sense. The first few days I would be in I would have to give at least two to three urine samples with no explanation. Now I had a reason for it! Also, I would get these bad back cramps that would go away after I finished my course of IV antibiotics. I had always assumed the cramps were just a back ache from sitting in the very uncomfortable chairs in the hospital or from being lazy at home, since I was a lot less mobile right before going inpatient. I also thought maybe they were phantom period cramps. Like when people have a limb removed they can feel pain in the limb even though it is no longer there. The cramping would happen when I wasn't sick too but would go away after a while, or I stopped noticing — I am not really sure anymore! But apparently they had to do with the UTIs I was constantly getting.

Much to my PCP's chagrin, every time I had the back cramps from that point on, I called the office and gave a sample. I wanted to test my theory. It turned out to be correct. The ONLY symptom I would get was the back cramping (besides the time in March 2015). And these were happening every few weeks — you can see why I was thinking phantom period cramps.

That is a lot of Bactrim to be on consistently. So I decided to look into alternative treatments. Since cranberry is widely known to be good for UTIs, I decided to give it a try. But because I have non-insulin-dependent CFRD, I didn't want to be drinking a ton of cranberry juice. Plus, I get mouth ulcers and didn't want to trade one issue for another. Instead I bought cranberry supplements. I now get them from CVS and they are the 4,200 mg soft gels. I take one twice-a-day. Since I started back in May, I have had one UTI and that occurred when I forgot to get more cranberry soft gels and ran out for three days. Three days was all it took to get another one. There have been a few times when I felt a slight twinge in my lower back and I bumped up the cranberry to two tabs twice-a-day for a few days. The pain went away.

I am very thankful that the cranberry is working so well for me. And since it isn't a medication, I don't feel like I am adding more toxins into my already overloaded body. For me this alternative therapy is working like a charm. ▲

> "Because I have non-insulin-dependent CFRD, I didn't want to be drinking a ton of cranberry juice. Instead I bought cranberry supplements."

AMY BRAID

Amy is 36 and has CF. She is a Director of USACFA. Her contact information is on page 2.

A Spoonful Of Honey To Keep The Doctor Away?

By Ella Balasa

Since I was young, my parents always have been advocates of natural therapies and supplements in addition to prescribed CF treatments. More recently, I have taken it upon myself to supplement my health routine daily by taking a spoonful of Manuka honey and washing it down with a half cup of water that contains a teaspoon of apple cider vinegar, first thing in the morning. I started this about a year ago, and I cannot be certain whether it has helped my overall health, but I don't believe it has had any detrimental effects.

Manuka honey is produced by bees that pollinate the Manuka bush, which is native to New Zealand. Manuka honey has a higher quantity of antimicrobial properties than honey made from any other plant species. Hydrogen peroxide and methylglyoxal are found in higher concentration in Manuka honey, which contributes to the antibacterial affect.

There have been studies conducted at various universities demonstrating this type of honey's ability to kill many types of microbes. Manuka honey has been used throughout history in wound care and is used in healing ointments today. It has not been shown to create resistance in microbes, unlike traditional antibiotics. Microorganisms mutate to develop mechanisms to resist the effects of traditional antibiotics. As we all know, uncontrolled growth of bacteria in the lungs, in cystic fibrosis patients, causes scarring and advances lung disease. Killing the bacteria is key to maintaining lung function and overall health.

ELLA BALASA

Apple cider vinegar is made from fermenting the sugars from apples, which creates acetic acid and this is the main compound in apple cider vinegar. Acetic acid can reduce bacteria counts and it has been used as a disinfectant and a preservative for this reason. Many CF patients have CF-related diabetes and apple cider vinegar has been shown to lower blood sugar levels. I have been pre-diabetic for a few years, but my sugar levels have not consistently stayed at a level where I would require insulin. I have been fortunate in this respect.

Overall, I believe a healthy, balanced diet, supplemented with a few vitamins and natural substances can have some health benefits. There is no cure for CF, so it is important to keep in mind that many treatments may subtly help fight this illness and augment your healthiness, but there is no single trick that will make everything better. ▲

Ella is 24 and has CF. She is a Director of USACFA. Her contact information is on page 2.

TILLMAN continued from page 15

(Cl-) ions across epithelial tissues. In patients with CF, the efficient migration of chloride ions is impaired. The research provides new insights into the proper functioning of the chloride ion gateway and mechanisms underlying its dysfunction in patients with CF. A better understanding of the structure and function of the ion gateway reveals how estrogen might interfere with the KCNE3-KCNQ1 channel complex, apparently worsening the effects of CF lung disease and leading to higher mortality rates and shorter lifespan for female patients. It is believed that proper functioning of the KCNQ1 channel helps mitigate fluid buildup in the lungs. When inhibited by high concentrations of estrogen, ion flow is impaired, allowing bacterial infections to take hold.

http://tinyurl.com/zrpgzpq

AND

http://tinyurl.com/hz437xa

Chronic CF Lung Infections Treated with Antibiotic Combination

A combination therapy with inhaled Bramitob (tobramycin) and Colistin (colistimethate sodium) improves lung function and respiratory symptoms in cystic fibrosis patients.

Continued on page 25

Reiki And Its Benefits

By Andrea Eisenman

When I think back to my first Reiki healing session, I realize that it has completely changed my life and health. I was headed to my acupuncturist's by subway, already frazzled because I was running late and worried about the health of two of my friends with cystic fibrosis and my mom. Of my CF friends, one was very ill and one was dying. My mom has COPD and has many other health-related-issues, too long to list here. My head still spinning, I walked in to my acupuncturist's office and, right away, Jackie, my acupuncturist, could tell I was an anxious mental mess. Before her treatment, she recommended I have a Reiki session with one of her massage therapists who is a Reiki master.

I didn't think that Reiki could possibly make me feel any better. As I lay on the massage table fully clothed, I just wanted to feel less stressed out and reduce my already frantic heart rate. Michael, the Reiki master, lightly placed his hands over specific areas of my head and body. After a few hand placements, I started to relax, my breathing slowed and I felt almost in a meditative state. I was not obsessing about my friends or my mom. My head started to clear. I was just "being." Or, as I would later learn, feeling present. I think the whole treatment lasted about 15-20 minutes and I felt relaxed and almost ready to nap. I felt like he had hit a reset button on me. I just felt much more at peace.

After he finished my treatment, I remarked on how much better I felt and inquired how he learned Reiki and whether anyone could. He said, yes, anyone can learn it, just Google Reiki instruction. So, that is what I did almost immediately. I signed up for level one with a woman who is a Reiki master about ten blocks from my apartment. Sometimes convenience is key.

The class was a three-day course and reasonable in price. She had written a book and requested we read it so we would know what to expect and have some information before embarking on her class.

ANDREA EISENMAN

I was excited because I loved my experience with Michael. As I started to read the teacher's book, I learned where Reiki had come from and how it was introduced to America. It talked about what the benefits are and even used a person with CF as a case study. It listed the hand placements for treatments for self-practice and for treating others. I was ready to learn so I could treat myself and friends, if they let me.

After learning level one, I was able to practice Reiki on close friends and myself. My teacher recommended treating myself daily and said that, with practice, I would deepen my abilities. Initially, I would practice Reiki and be very aware of everything – the time, my hands in the positions, my breathing and sometimes feeling bored. But soon enough, I started to enjoy how I was feeling – good.

Usually, my hands heated up or started to tingle in certain placements. It did feel meditative and relaxing. But Reiki didn't just make me feel relaxed, it helped with many things depending on that day and what I was feeling. If I had stomachache, once I placed my hands on my abdomen and felt my hands getting warm, my stomach would gurgle and usually pain was gone or lessened. My self-practice also eased my migraines and my sinus pain. When I placed my hands over my eyes and sinus area, I would sometimes feel a movement or the lining inside shrinking, especially when my hands would get hot. So I guess Reiki treatments were also reducing inflammation in my body. I have also experienced a reduction in pain in my hip and back after a self-treatment. I even felt my blood sugars were more controlled.

Six months after learning level one, I took level two. And that did start to truly deepen my practice. When I treated myself, I was falling asleep. It was a great way to take a break in the afternoon and would lead into my almost daily nap. I have even practiced when I feel I have a cold or sinus infection and have had some relief. Now I practice at night as I am going through peri-menopause. I do not sleep through the night, so if I wake up at 3 a.m., I will start the hand placements and usually fall back to sleep within an hour. This may sound like a long time but, without it, I can be up for two to three hours in middle of the night. Not enjoyable.

What is Reiki (pronounced ray-

Continued on page 32

THROUGH THE LOOKING GLASS

PHOTO BY STEPHEN BOYER

Vrksasana

Magnificent tree
reaching upward toward
the sky
grounding me

Miraculous breath
flowing throughout my body
sustaining me

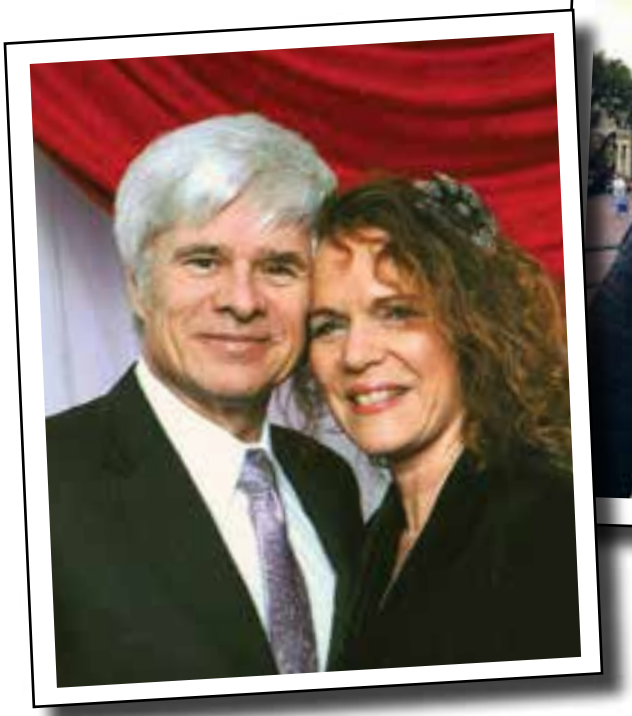
Mind, body, spirit
seeking balance and whole-
ness
strengthening me

-R. Petras, 2000

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

FROM OUR FAMILY PHOTO ALBUM...

CHRISTIAN PATON AND MICHELLE STILES IN FRONT OF BUCKINGHAM PALACE, ENGLAND.



VICTOR AND LINDA ROGGLI.

JEANIE HANLEY, CENTER, WITH HER FAMILY. FROM LEFT TO RIGHT: SON KEVIN, DAUGHTERS MARIA AND JESSIE AND JOHN, HER HUSBAND.

DR. CYNTHIA BROWN AND JEN WEBER AT A CF WALK.

BRIAUNA PETERS EXERCISING OUTDOORS.

ELLA BALASA IN IBIZA, SPAIN.

INSIDE DEVIN'S APARTMENT, CELEBRATING THANKSGIVING. FROM LEFT: HIS BROTHER BRENDAN, BRENDAN'S LOVER JENN, DEVIN, HIS MOM KATHLEEN, HIS GRANDMA PATRICIA, HIS GRANDFATHER MICHAEL AND HIS FATHER SCOTT.

ACTIVE FOR LIFE

Interview With Briauna Peters

By Aimee Lecointre

I've been friends with Brie for a few years now. We met through social media and were even able to meet in person at a friend's wedding. I've been following her story for a few years now and wanted to share it with you all. I think Brie's story is something many of us can relate to and is something that can inspire and motivate us all to take the step to incorporate activity into our treatment regimen.

Hey Brie! Thanks for letting me interview you to share a bit of your story with our community. First, can you share a little bit about yourself:

My name is Briauna Peters. I was diagnosed with CF at 10 months of age, at Lucille Packard Children's Hospital (otherwise known as Stanford) and have continued with treatment there ever since. I currently reside in far Northern California, in a little town named Ferndale, along with my husband. I have been married for three years and have two amazing stepchildren, Preston and Macie, who have given me the chance to be a momma. They are truly a blessing. My husband and I both were raised in the Redding, CA, area, where we still travel quite frequently and hope to get to move back to soon.

All right, let's get to it. When it comes to CF and exercise, has exercise always been a priority in your life?

Growing up with CF, my doctors always told my parents to keep me as involved in as many physical activities as I could. I grew up always going to dance class, cheerleading and singing. It wasn't until going off to college where it hit me how important staying active was. After high school, I wasn't involved in any type of daily physical activity to keep me in shape and, due to that, I ended up getting very sick and having to drop out of college spring semester of my sophomore year. At that point I spent about two years just figuring that that is how it was going to be and that I didn't have much control over my CF other than doing my regular treatments. Then it hit me, I needed to make a change!

So what motivated you to start making exercise a regular part of your routine?

I slowly saw my body deteriorating and I hated it. Doctors ended up placing a g-tube in me because I could not gain enough weight on my own. I was not getting anywhere and I was continually seeing numbers get lower. I was living but not truly living at my best. I knew without a doubt that nothing was going to change for the better unless I tried something new.

How has exercise impacted your health and life with CF?

I saw a change in my life when I started implementing exercise as part of my daily routine. Vitamin levels and blood work were normal due to changing my diet. I started gaining true weight for the first time ever. I had more energy and my lungs are more stable than they ever have been! It was after my first year of really getting back into exercising every day when it really hit me that the work is WORTH IT. I

BRIAUNA PETERS LIFTING WEIGHTS.

remember hitting my one-year mark of no hospitalizations. An amazing feeling! Doctors were impressed, to say the least, since previous to my change I was in every four to six months. Hitting that two-year mark makes this journey even better. Staying consistent with my healthy diet and physical fitness is what truly makes a difference in keeping the CF monster at bay.

What type of exercise do you typically do? What do you enjoy most?

I love weight training. No doubt it is my favorite type of exercise, no matter how simple it may seem. It has also proven to be the most effective type of exercise for me as well. I know how to feed my body correctly so it is powered properly so I gain muscles and not lose weight. Now, that doesn't mean I don't love dancing, hiking, wakeboarding, swimming or pretty much anything that is outdoors, but during those activities your body is exerting larger amounts of energy that requires a more extreme diet than I prefer to consume on a daily basis.

You exercise mainly at home. Do you feel you are still able to get in a productive workout? What are benefits you have noticed to working out at home?

Most definitely! 98% of my workouts are done in my living room. You don't need a lot of space, and you don't need tons of weights, especially when first starting out. Weight-bearing exercises, such as pushups and squats, can give you very successful workouts. Working out at home can be just as effective as going to a gym. I will visit a gym every now and then to get in a heavy lifting workout, but I tend to stay away for the most part due to the germ factor! Getting stares from coughing also isn't the most fun, but on the other hand, crushing a great workout and seeing people's faces while you do things they didn't think you could do while taking a meter dose inhaler in between reps is kind of rewarding! Anyway, I still prefer working out at home.

What keeps you motivated to stay active? If you ever get into a rut, and fall out of your routine, what helps you get back at it?

RESULTS. Nothing is better than seeing your hard work pay off. Fueling my body correctly and not feeling bloated, having daily energy after workouts, fitting into clothes that never used to fit and ultimately feeling like I can take a deeper breath after. That is why I choose to push every day for that bigger breath! Of course there will always come a time when I get thrown down, am tired or get sick. There is a time to rest. It is important. But there is also a time to make sure you don't get stuck in that rut. That is where you need to push yourself every day to get up and do something active, even if you can do only 10 squats! Losing all your progress will suck a lot more than trying for just 10 minutes, I promise. To get myself motivated, I try to visualize myself "healthy," what my goals are and keep that positive image in my mind. Your mind is so powerful in what you can and cannot do!

Any words of advice for those struggling to start exercising or just being active?

Believe in your future self. No matter where you are right now, there is a way that you can start getting active, no matter how small the steps. The obvious ones are changing your diet and choosing the healthier snack and fueling your body correctly throughout the day. The second is implementing some sort of exercise routine. A lot of people will go full bore their first day and wind up getting sick or discouraged because they over-exerted themselves. The key is taking small steps. Whether it be a squat challenge, lifting weights, increasing how far you can walk each day, start simple, start slow and work your way to your goals. No one, and I mean NO ONE, is going to be a gold medalist, superstar athlete overnight.

Through your experience with CF and exercise, you have created a foundation called *Muscles To Breathe*. Can you briefly tell us what your mission with M2B is?

Muscles To Breathe is my passion. My mission for *Muscles To Breathe* is to make nutrition and fitness products readily available to the CF community while bringing awareness to what life is like with cystic fibrosis. Throughout my own fitness journey, I have learned first-hand the financial struggles in order to maintain this healthy lifestyle, especially on top of taking care of my CF. Too many CF patients can't afford fitness essentials like nutrition supplements, fitness products and gear on top of insurance costs, hospital "tune-ups" and mainstream treatment therapies. Through my non-profit, I hope that I can help others like myself to thrive and live their lives happy and healthy! *Muscles To Breathe* is currently in the early stages of Section 501(c)(3) business entity application and hopes to receive its IRS title soon.

Thank you, Brie, for taking the time to chat and sharing your story with us! ▲

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

PARENTING

The Power Of Playing

By Dana Giacci Rogers

This issue of *CF Roundtable* is dear to my heart because non-traditional medicine is something that I have spent a lot of time on this past year. I would like to take this opportunity to talk a bit about a form of alternative medicine that I use and how it relates to my parenting style living with CF. Outside of being a mom of three, I am also a registered yoga instructor and have recently received my certification in teaching children's yoga. In recent years, yoga has become more and more mainstream. The yoga community only continues to grow, but what is it?

In traditional yoga teaching, yoga is the practice of connecting the mind, body and spirit of an individual for a more wholesome outlook on well-being and for some spirituality. There are many different types of yoga that I feel the CF community can benefit from. This includes *asana* practice, which is the movement, strengthening and conditioning that most have experienced in a yoga class either at a gym or in yoga studios. There is also *pranayama* practice, which includes a myriad of breathing exercises that focuses on breath expansion and retention, which not only aids in clearing secretions but also in respiratory endurance for more demanding activities. Finally, there are many limbs of yoga that some might group together and classify as simply "meditation," which can benefit many things including anxiety, focus and self-discipline when it comes to treatments, med schedules or time for oneself. There is something for everyone in a yoga practice. It does not need to be a spiritual practice unless the practitioner is searching for it to be so. It does not need to be a physical practice for the same reasons.

How is this applicable to CF and parenting? Yoga is an activity that I have chosen to be an addition to my typical treatment routine. However, as I practiced at home, I watched my three-year-old try to keep up with me. Soon, we went from her standing under my feet to finding her way to her own mat. She and I now do a yoga practice together. Once I realized her enjoyment of what she calls "playing yoga" with me, I spent the extra time and effort so that I could include my children in my activity if they were interested.

Which brings me to the point of my column today. The power of playing with our children. As parents with CF and as parents of children with CF, the more we baby ourselves or find excuses for ourselves because we feel tired or breathless, the worse those feelings can become. If nothing else we need to allow our children to encourage our well-being by simply being active with them, which in turn encourages their own active lifestyle. My three-year-old practices yoga with me and it is in those moments as a CF parent that I realized being active together is our way of playing; and the more I focused on playing, the more I realized I just needed to count it as an additional treatment time.

> *"My three-year-old practices yoga with me and it is in those moments as a CF parent that I realized being active together is our way of playing."*

DANA GIACCI ROGERS

So often in CF parenting, it might feel as though treatment times and caring for our kids are at odds with each other. But not when we are playing. Yes, in my house it started as yoga, but it expanded little by little. Once I realized this, it was easy to take it one step further beyond our yoga mats. My daughter loves to play soccer, and tag. I cannot express to my CF community how much I dreaded those two activities as a young child. However, now they are an opportunity—an opportunity for an extra treatment, an opportunity to love my kids, an opportunity to be a better parent, an opportunity

for me to not feel like a CF patient.

So this is my encouragement to our broader CF community **and** to those parents with CF kids. If there is an activity that you love that you can include them in, then do it! Yoga, running, basketball, tennis, baseball, soccer—go play! Even if it feels like a little bit of work. Go run around and play tag. Set a timer if it is hard to get started. Perhaps start at 10 minutes, and then go up to 20 and so on. It's good for us, and so, so good for them. They don't care if you need to stop and cough, and they don't care if you need to take a breather. Get up and go together. This goes beyond CF advice. This is as good as alternative medicine can get. Good for all hearts involved. GO PLAY!

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

Please remember, if you have any questions regarding CF and pregnancy or parenting, e-mail me and I will do my best to address those questions in the next issue's column.

Stay Healthy,
Dana ▲

TILLMAN continued from page 17

Researchers conducted an exploratory study to assess the efficacy of Bramitob (known as Bethkis in the U.S.) combined with Colistin on lung function in CF. They recruited 41 CF patients, ages 6 to 50, who have had chronic P. aeruginosa infections for six months. Participants received Bramitob inhaled twice a day at 12-hour intervals for 28 days, followed by Colistin inhaled twice a day for 28 days. This cycle was repeated for six months.

Results showed that both mucus production and coughing decreased by 39% in patients treated sequentially with Bramitob and Colistin. In addition, FEV_1 absolute (the volume of air that can forcibly be blown out in one second after full inspiration; a measure of lung function) increased by 4.9%, and FEV_1 relative to baseline increased by 9.1%. The authors concluded that treatment with sequentially combined inhaled antibiotics is very well-tolerated and may have a beneficial effect on lung function and respiratory symptoms.
http://tinyurl.com/hpjazke

New cystic fibrosis device to reduce antibiotic usage

A medical device for cystic fibrosis sufferers which lessens their exposure to infections resulting in fewer doctors' visits, reduced antibiotic usage and shorter hospital stays, is expected to be market ready within two years. University of Limerick inventors recently secured funding for the new percussion device that helps remove mucus from the airways. Currently, there are percussion-based chest physiotherapy devices on the market, but these can sometimes become reservoirs for the bacteria that cause infections in CF patients. Because the new device, SoloPep, is disposable, it poses no threat of reinfection.
http://tinyurl.com/gpe6qjt

Risk Factors for Severe CF Lung Infections Caused by S. aureus Identified

Several factors may worsen lung infections caused by the bacteria Staphylococcus aureus in cystic fibrosis patients, from bacterial load to co-infections, according to a new study.

Researchers enrolled CF patients with chronic S. aureus airway presence as part of a multi-center study. The team assessed lung function taking into account several parameters, such as S. aureus bacterial density in sputum and throat samples, co-infection with other CF-associated pathogens, presence of S. aureus in the nasal tissue, clinical status, antibiotic therapy and the levels of IL-6 and IgG-levels, both of which serve as measures of the degree of immune response against S. aureus infection. Results indicated that the age at which patients became infected with S. aureus influenced the amount of bacteria present in sputum samples (but not throat samples). That is, older patients had significantly higher bacterial loads in the sputum than younger patients. Bacterial density in throat samples (but not sputum samples) also significantly correlated with a decline in lung function each year. Loss of lung function was also associated with other parameters, such as exacerbations, presence of small-colony variants of S. aureus and co-infection with the bacteria Stenotrophomonas maltophilia. Patients with increased blood levels of IL-6 had higher density of S. aureus in their sputum and also showed lung function decline over time. Those with nasal cultures of S. aureus had better lung function over time.
http://tinyurl.com/zuxgyrb

CF Pseudomonas Infection Best Treated by Cayston, Comparison Study Finds

Cayston (aztreonam) was deemed the best treatment across a range of parameters in a study comparing inhaled antibiotic treatments against Pseudomonas aeruginosa lung infections in patients with cystic fibrosis. But the study also demonstrated that Quinsair (levofloxacin inhalation solution) came in as a good second, improving lung function better than Cayston in the long run.

Continued on page 29

WELLNESS

What Do You Call Meditating While Vesting?

By Julie Desch

There are two types of people in the world: those who hate candy corn and those who cannot stop eating it. I fall into the latter group and as I enjoy my sugary delight, obsessively eating each differently colored section one at a time, piece by piece, it strikes me that people seem to fall into the same pattern when it comes to meditation. I clearly am a big fan, yet my life partner visibly recoils at the idea of following the breath for more than an instant.

With that in mind, this article is directed at those of you who loathe the idea of a meditation habit or who just simply believe you are incapable of the practice. Come to think of it, it's also for those who think they have no time to meditate. I used to wander in and out of this last group, until I discovered a "no-brainer" way to incorporate a meditation practice into my day.

I don't know why it took me so long to invent the practice of "medishaking" (I'm working on the patent). It seems now that being physically tethered to a device for 30 minutes at a time would be an obvious solution to the problem of "not having 30 minutes/day free" to meditate. The fact that it took me approximately 15 years to see an indisputable resolution to my lack-of-time issue is somewhat embarrassing to admit.

At the risk of extending the candy corn metaphor too far, there are three distinct benefits to meditating while Vesting. First, when trying to establish a new habit, every source tells you to "attach" the new habit to one that is already established. A good example of this is when I began to drink a full glass of water (new habit I was trying to institute) while waiting for my coffee to brew (*extremely* well-established habit). Only a tiny amount of will power was required to do this (and maybe a yellow sticky note on the coffee maker as a reminder). Before long, I felt weird if I didn't drink water first thing in the morning.

Vesting is a habit that I trust we all have well established by now. If you are like me before my medishaking epiphany, you occupy that time browsing on your phone or computer, watching television or Netflix or doing some other less-than-important thing. What if you spent just a portion of that 30 minutes calmly practicing breath awareness? Or if breath awareness isn't your thing (and this is perfectly fine), you could spend a few minutes following sensations of the body as it wears an airway clearance device. The Vest makes either option work very well, which I discuss below.

It isn't necessary to commit to the full 30 minutes at first. In fact, I would suggest starting with just a few minutes. If your Vest is like mine, it is programmed to shake me at a certain frequency and intensity for five minutes, then stop to allow for coughing for one minute, then proceed for six cycles in total. So you could commit to five minutes at first and then, if you are so inspired, extend the time as you see fit.

> *"We live so much of our lives in our heads, it's easy to become almost disconnected from our bodies."*

JULIE DESCH, MD

The second benefit to using Vest sessions as your meditation time is that the physical sensations created by strapping a jackhammer to your torso amplify the usual sensations of the breath or body. For me, this amplification makes staying with sensations easier to do. When I am following the breath while wearing the Vest, the shaking intensity and even location feels very different on the in breath as compared to the out breath. At each frequency, this changes in interesting ways. Curiously, my nostrils resonate for some reason with the 18 Hz frequency, but only on the in breath. It's weird...and sort of fun to watch. Body sensations become more prominent and interesting as well. Sometimes, beginning meditators have a hard time actually feeling sensations of the body. We live so much of our lives in our heads,

it's easy to become almost disconnected from our bodies. But the Vest certainly fixes that problem. The prominent shaking of the torso is certainly easy to be aware of and is a great anchor to use in meditation.

Finally, just as the tiny white tip of the piece of candy corn is almost singularly reason to live, the minute of stillness breaks between the five-minute-long shake cycles is, to me, a very pleasant collection of moments. I find that this minute makes all of the times I have to "gently bring my awareness back to the breath" completely worth it. When the body stops shaking suddenly, it almost feels like my body is in free-fall, relaxing into a sinking feeling that is accompanied by tingling and vibration through my whole body. It's hard to describe, but definitely worth checking out! ▲

Julie is 56 and is a physician who has CF. You may reach her at jdesch@usacfa.org.

In Memory

Talana Lyda Fairfax, 34
Cottage Grove, OR
Died on August 31, 2016

Anne Childs Williman, 63
Middletown, OH
Died on September 21, 2016

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

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

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DIAMOND SUSTAINING PARTNERS

IN MEMORY OF
LAUREN
MELISSA KELLY

SEARCHING FOR THE CURE

Many Hopeful Options Abound At The NACFC

By Reid D'Amico

Walking through the North American Cystic Fibrosis Conference left me with a big smile as I saw that companies are tirelessly working to give those with CF a normal life. With the introduction of CFTR therapies, and the possibility of gene therapy, the community finds itself more optimistic than ever. However, it is still important to take a step back and appreciate the therapies that are entering clinical trials that do not directly target CFTR. In fact, we should embrace them twofold: because those with rare CF mutations are still waiting for their CFTR modulators, and those who are fortunate enough to have CFTR therapies may find improvements in health by replacing or adding additional therapies to their regimens.

CF therapies are a multi-pronged effort to protect tissues from damage and alleviate symptoms. CF patients struggle with pancreatic insufficiency, chronic infection, inflammation and dehydrated airways. All too well, we understand that each of these symptomatic classifications of CF requires a different medication or therapy. However, some of us are now realizing that these new CFTR medications may protect us from some CF manifestations, but they may not allow for us to totally stop our other therapies. This is why we must continue to embrace the therapies that enter clinical trials that are not just CFTR modulators.

For example, there are new antibiotic treatments, next generation anti-infectives, new medications that address inflammation, devices and medications to help digestion, new CFTR medications, and treatments to aid mucocilliary clearance all in or entering clinical trials.

In CF, we have all learned that our disease is rooted in a defective protein. However, we now know that some of the problems seen in CF are not just related to the CFTR protein's defect. Research has recently found that deranged ion levels may be related to other ion channels other than CFTR. CFTR is not be the entire story when it comes to treating cystic fibrosis. And up and coming research may show us that we may find better luck treating proteins that are found in the same molecular pathway as CFTR.

REID D'AMICO

Growing up, life expectancies for CF were low, and I remember being so inspired anytime I saw someone with CF in the public eye. The past few years have been ones of tremendous growth for our community. We now see that CF has become a king in rare disease, and has outlined plans for other foundations and researchers to follow. We will continue to live longer with an increased quality of life. But we cannot forget how we got this far: our appreciation for clinical trials. CFTR therapies are groundbreaking—there are no questions about that. However, we must not lose sight of our final goal: a healthy life for *all* with CF. It is because of this over-arching vision that we must continue to work for better treatments for all with CF. Therapies that do not address CFTR are therapies for the entire community, including those of us with mutations that can now be directly treated. ▲

Reid is 23 and has CF. He is a Director of USACFA. His contact information is on page 2.

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BYRNES continued from page 7

save energy for other life activities.

Let's look at the spiritual benefits of healthy willpower. What if we had the best willpower ever, if we sleep, exercise and eat right, do our treatments and do all the right things to set and accomplish our goals? The bigger question is "Why?" We need a *reason* to have willpower. We need a spiritual purpose for this enormous effort. Maybe it's for our loved ones, for the CF community, for God, to respect this one and only life we have. Nietzsche said, "He who has a *why* can bear with almost any *how*." With a greater purpose, we can expend the effort to endure and survive.

Goals can sometimes be a "map" that will guide us toward our God-given purpose. Without goals, we may wander aimlessly not knowing our direction. Some people think goal-setting is unspiritual because it shows a lack of trust that God is leading us to what we are meant to do. I feel like setting goals is a way to use our God-given gifts and talents to do good work and fulfill our God-given potential. God wants our active cooperation in fulfilling our fate (1 Cor. 9:24-27).

Fulfilling goals is due to willpower, intention and having time and energy. But fulfilling or not fulfilling goals is sometimes out of our control. For many years, I'd say, "It would be nice to fulfill x, but if I can't, it's okay too." When I was able to get my LCSW, I felt like there was divine intervention. To be this healthy, to land a perfect job, to find the right combination of clinical supervision, to have the funds to take prerequisite courses and have the time to do this, all aligned so perfectly. Sometimes goal-making is like that. We have to wait patiently, very patiently, for the right time.

I encourage you to practice McGonigal's willpower traits. Set goals that motivate you. See what's waiting for you. ▲

Isa is 44 and has CF. She and her husband, Andrew, live in Redwood City, CA. You may contact her at: Isabear27@hotmail.com.

TILLMAN continued from page 25

http://tinyurl.com/z4banne

Positive Data Presented in Cystic Fibrosis Patients Using Quinsair

Raptor Pharmaceuticals recently presented the results of a meta-analysis comparing the effectiveness of inhaled antibiotics for cystic fibrosis (CF) patients and Pseudomonas aeruginosa lung infections. The results showed that the company's levofloxacin inhalation solution Quinsair was comparable in effectiveness to three other European-approved inhalable antibiotics. The meta-analysis included all randomized clinical trials assessing the use of inhaled antibiotics in CF patients with follow-ups of four or 24 weeks. It compared the effectiveness of aztreonam, tobramycin, colistimethate sodium and Quinsair against lung infections involving P. aeruginosa. Treatment with aztreonam (75 mg) three times daily was found to result in the greatest increase in forced expiratory volume in 1 second at four weeks. But Quinsair (levofloxacin) was found to be superior to colistimethate sodium, tobramycin inhaled powder and tobramycin inhaled solution. The results showed there was no evidence indicating that the other solutions were more effective than Quinsair. Results from the 24-week analysis demonstrated that the inhalation solution Quinsair was linked to the lowest hospitalization risk, with a 96.5 percent of probability to be the most appropriate treatment option.

http://tinyurl.com/grfpn98

Calcium induces chronic lung infections

During disease progression, P. aeruginosa adapts its strategy by switching from acute to chronic virulence. It stops the production of virulence factors, such as bacterial injection apparatus and toxins and, instead, produces a protective matrix and reduces its growth rate. The environmental signals directing this transition are so far unknown. Researchers have now identified calcium as a signal that specifically triggers the switch to chronic virulence. In Pseudomonas, a central signaling pathway senses environmental information and ultimately determines whether the pathogen will undergo the acute to chronic virulence switch. Although the components of this pathway are well-known, none of the external signals modulating the switch are defined. The researchers have now discovered that a receptor located in the bacterial cell envelope monitors the calcium concentration in the environment and transmits this signal into the cell. Elevated calcium levels trigger the switch to a chronic program: The bacteria protect themselves within a biofilm structure, reduce their growth rate and by that increase their drug tolerance and persistence. Most of the isolates from airways of CF patients have retained their calcium sensitivity. This allows these bacteria to constantly adapt their virulence in response to the often changing conditions in the airways. One of the characteristics of CF is deregulated calcium homeostasis. The researchers assume that elevated calcium levels in patients promote the switch from an acute to a chronic state of infection. This is of advantage for the pathogen, as it may ensure its long-term survival in the respiratory tract.

http://tinyurl.com/zfn3gxg

Catabasis Pharmaceuticals to Present

Continued on page 32

Being Your Best Advocate

By Jeanie Hanley, M.D.

As a physician and adult with cystic fibrosis with many hospitalizations under my belt, I've advocated for others and myself for over 30 years. I've watched medicine morph from the "paternalistic" age to the information age. During the paternalistic age, patients trusted the information they received regarding diagnosis, treatment, course of the illness and prognosis. Doctors also spent a lot more time with patients back then. Even so, this was the "Yes, doctor," age as opposed to our current information age, or the "What about this?" or "Why that?" phase, which has ushered in very informed individuals. As a result, we have been able to become partners in our care and more engaged due to easily accessible research of symptoms, side effects, drug interactions and more.

My first advocate and the one who questioned everything at every visit was my mother. I was my own second advocate after fending for myself once I attended college at 18 years old. Self-advocacy never ends and isn't always easy. Having other advocates relieves considerable stress. My best advocate has been my husband and others who step in during hospital stays and when I'm just too sick and/or tired to deal with pharmacies, insurance companies or even clinic visits.

Over the years some basic principles of advocacy have become apparent. First, have a friend, significant other or family member be aware of your care from the very beginning. If you wait until health goes south, then advocacy will be much more difficult for you and everyone involved. Second, when health issues arise, enlist help as early as possible. Again, involvement of your advocate works best when begun at the start of a flare-up. Third, build your medical resources—including people and places who you know are especially responsive to your medical needs, e.g. a particular member of the healthcare team—a nurse, personal physician, physicians in different specialties, pharmacist, social worker and preferred urgent care centers or hospitals. Having a list of these resources readily available enables you and others to get a handle on who or where to turn when emergencies arise.

JEANIE HANLEY

To advocate for yourself or others, preparation is the mantra. That includes writing down questions for each health visit with a doctor, nurse, nutritionist, pharmacist, social worker and/or whoever else will be seen that day. And I mean actually writing or typing the questions out. Doing so helps to organize your thoughts by noting the sequence of events that led up to the current problem and zero in on the most pertinent details and questions. I can't stress how important that is. If you also educate yourself and investigate your symptoms, medicines and diagnosis, then the questions can be better tailored by incorporating what you've learned.

Education via Internet can be tricky sometimes if you consult a source that may not be particularly accurate or someone who has put two and two together and has arrived at three. For this reason, I've compiled my own list of reliable Internet sites that I'm confident will be accurate. These include MayoClinic.org, UpToDate.com, MedlinePlus.gov, Healthfinder.gov and other health hotlines. For good measure I suggest double-checking the Consumer and Patient Health Information Section (CAPHIS), which lists the "Top 100 Health Sites You Can Trust" at http://caphis.mlanet.org/consumer.

Education through others can also be informative. I am careful to note variations in our symptoms and care (e.g. different phenotype or genotype), which may result in different responses. For example, since starting Kalydeco, my energy has soared to the point of insomnia. Talking with others about their experiences with Kalydeco certainly gave me food for thought and added to my question list, especially with a relatively new medication where all the side effects may not be known. But no one had experienced insomnia as a side effect, nor was it listed in any drug resource as a side effect. Insomnia is most likely an unrelated issue. But my CF clinic and my husband are aware so if any new association arises in the future, we may gain insight together.

Along with preparation and education is something I've dubbed "The

Observer Effect," which is the simplest form of advocacy. The Observer Effect describes the benefits gained when someone accompanies you to your clinic visits. My absolute criteria for the "observer" is that they should be at least 18 years old and will make eye contact and listen to all healthcare personnel who enter the clinic room. Secondary criteria that would be very helpful are they are confident enough to ask questions and will take notes, especially since studies show that 40 – 80% of information presented during a clinic visit is forgotten immediately or remembered inaccurately.

The first time I realized "the Observer Effect" was during a follow-up visit when my 18-year-old daughter accompanied me, not for advocacy, but just to hang out with her mom. I had no pressing concerns, but I was surprised by the extra time and attention I received by just having her with me. I knew she wasn't quite listening, since she had other 18-year-old stuff on her mind, but she still made eye contact and appeared to listen, even though I knew her mind was a mile away. Yet, here the doctor was spending much more time than usual. Looking back, I've similarly spent more time with patients who brought family with them, making sure that all understand. But I did not carry that experience over to my own care. Now I do and bring my "observer" as often as possible and recommend that others do the same.

Another form of self-advocacy is scheduling and keeping appointments, even when well, which helps to maintain your health and fine-tune care. Sometimes, I have had thoughts of skipping out on appointments because I'm feeling relatively well. But then I remind myself (or my husband does) how those appointments end up being the most informative because I'm more alert and have more stamina to ask questions that may improve my care in some way, either with greater understanding, new research or a simplified regimen. When I'm well, the staff seems to respond more energetically, too.

Knowing your medications and sharing them with advocates, along with therapy routines, is key. Even more important is to use the medications every day with the frequency needed each day whether it is once daily to four times a day or more. Setting up a routine and working it into a daily schedule is difficult at first when new meds are added or the frequency is increased. Once a routine gets going, then the difficulty subsides.

> "Self-advocacy never ends and isn't always easy. Having other advocates relieves considerable stress."

Lastly, know your insurance policy. This is boring and time-consuming but a necessary aspect of your health. We all should learn what is covered and what is not, regarding medications and their prices; services, and hospitalizations, and where we can go for care and how much we'll pay when we get there. When the bill arrives, scrutinize it for the charge, by whom and what level of care. I've found numerous errors. A physical therapy clinic and an ENT doctor both billed my insurance as a "new visit" even though I was a regular patient. As a result, my copayment was much higher than it should have been. Informing both reduced my bill to a fraction of the original amount. Hospitals charged for services that weren't given. One clinic sent the bill in a year later and my insurance wouldn't cover it. When the clinic asked me to pay in full, I refused since it was their responsibility to submit to my insurance within the year. I provided the EOB (Explanation of Benefits) I'd received to prove it. I never heard from them again. I believe you get the point.

As individuals with a chronic illness, we deserve to be heard and treated with dignity and respect. By being engaged and involved in our care, we promote our own health. We can stop a problem in its tracks by prevention—staying on top of our medication regimen, exercising, understanding our treatments, educating ourselves and following up regularly. We can enter clinical trials (see "Searching For The Cure" on page 28) and join support groups online or be involved in CF organizations. We can get legal help through the CF Legal Hotline (see "Ask The Attorney" on page 4). Of course we can read *CF Roundtable* publications, which offer support to us all, our advocates and healthcare providers.

The benefits of advocacy on our illness are numerous—reduced stress and improved coping skills and quality of life. With successful advocacy comes empowerment within our advocates and us by increasing our awareness of options and knowing how to proceed when health takes a downturn. And guess what? Taking charge of our health and giving ourselves options leads to a greater sense of peace and happiness. Sounds good to me. ▲

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

EISENMAN continued from page 18

key), you might wonder? It is a balancing modality. It can be unique to the individual or based on what the individual is going through. It is not necessarily one thing defined by this or that. I have practiced on friends and with my mom. All of them have had various problems or aches and wanted relief. Sometimes they will fall asleep or just feel better overall after a treatment. Sometimes they will see colors and tell me all sorts of things that they are feeling while lying on the massage table. They will say: "Your hands are hot, or so cold." They will ask what I am doing and I reply: "I am just placing my hands on various parts of your body." Once I learned Reiki, I just got out of the way and let the healing begin. I am not usually thinking anything, thoughts come and go, like when one meditates.

Now, don't try this at home, kids. Well, you can, if you find a book that gives you hand positions, but one really needs to learn from a Reiki master with hands-on instruction and practice. Once I had my instructor's book, but before class, I tried using the hand positions therein and felt next to nothing except my hands on my body. Nothing like Michael's skilled Reiki treatment. But once I took the level one class and was initiated by my teacher through meditation, it was then that I felt the difference in my hands. My teacher said we now had "Reiki hands" and it did feel that way. When I placed my hands on my body, they did feel different than before I joined her class.

I know it sounds odd and woo-woo. But let me say that, had I not experienced Reiki healing myself, I would be on the skeptical side of the fence right along with you. I have found that I am sick less often and my mental state is better than before I was using Reiki.

I have been practicing now for just over five years. I go to Reiki clinics to practice on others and receive a treatment, too. I even found a Reiki circle near my mom's house in Long Island. I still practice on myself daily. And about a year ago, I became a Reiki master, which has allowed me to deepen my practice further.

One very important aspect about Reiki healing for me is I can practice on myself. So I can treat myself and not rely on others as I do for acupuncture, massage, any of my usual medical procedures and treatments. It is quite empowering, especially since I have experienced the benefits that I have. I think that is the reason I am less depressed and hopeless. CF, as we all know, can be daunting, even with a lung transplant. I still have sinus problems and at times my gut goes out of whack. Reiki helps me cope and feel better.

My hope is that if you have the opportunity to try it, you will check it out. You might enjoy it and it might make a difference in your health, as it did for me. Or, learn to practice Reiki and open up a new world of possibilities for yourself and those lucky friends whom you will treat. ▲

Andrea is 52 and has CF. She is a Director of USACFA and Executive Editor/Webmaster of CF Roundtable. *Her contact information is on page 2.*

TILLMAN continued from page 29

CAT-5571, a Novel Activator of Autophagy, as a Potential Treatment for Cystic Fibrosis at the 30th Annual North American Cystic Fibrosis Conference

Catabasis is developing CAT-5571 as a potential unique oral treatment for cystic fibrosis (CF) with potential effects on both the cystic fibrosis transmembrane conductance regulator (CFTR) and on the antibacterial clearance of Pseudomonas aeruginosa. CAT-5571 is a small molecule that activates autophagy, a process that maintains cellular homeostasis and host defense mechanisms, and is known to be impaired in CF. Catabasis has shown that CAT-5571, in combination with lumacaftor/ivacaftor, enhances cell-surface trafficking and function of CFTR with the F508del mutation. Catabasis has also shown that CAT-5571 enhances the clearance of Pseudomonas aeruginosa infection in preclinical models of CF, irrespective of CFTR mutation status.
http://tinyurl.com/gqsuqq3

Mold Toxins in Nuts and Corn Can Aggravate CF Symptoms, Cause Infections

Toxins produced by mold found on nuts and corn can exacerbate cystic fibrosis (CF) and pave the way for airway infections, as they block self-protective mechanisms for the lungs. The study showed that the toxins affected the movement of airway cilia — the hair-like structures that clear dust and microbes from the airways — effectively preventing the cleaning of the airways.

The toxins are called aflatoxins and are already known to cause cancer and liver damage. Using human airway cells grown in the lab, the research team discovered that the toxins block the movement of airway cilia. This mat of tiny "hair" lining our airways constantly beats to clear them of dust, mucus and microbes. When the system is not working as it should, microbes have an open door to the airways. But the researchers' work also has an upside. They discovered that the toxins activate an enzyme called PKC (protein kinase C) to arrest the movement of cilia. Drugs blocking PKC are already being explored for other conditions, and when the team tested two such experimental drugs,

Pay It Forward

BRONZE

Michelle Allen (in honor of my 65th birthday and our 20th anniversay)
Shirley Althaus (in memory of son, Stewart Kessinger & daughter, Janice Kessinger)
Anonymous (in memory of Kathy Estabrook & Tom Desch)
Susan Baldwin, M.D. M.P.H. (in honor of Andrea Eisenman & Jeanie Hanley)
Bridget Barnes (in honor of Lucy Barnes)
Rick Birkner
Ann Black
Joan Finnegan Brooks
Karen Burgess
Lisa Cissell (in memory of sister, Mindy Filiatreau)
Nancy & Robert Coleman (in honor of Claire Coleman Herriot, 17 years post-transplant)
Kevin Corr
Alan Cunningham
Andrea Eisenman (in memory of Anne Williman)
Mary Lou Figley
Casey Forbes
Shelly Forbes (in honor of Michelle Stewart)
Tom Gallagher (in honor of Colleen Black
Jeanie & John Hanley (in memory of Theresa Ponaman Boujie)
Jeanie & John Hanley (in honor of Mary Ellen Sampson)
Marie F. Henry (in honor of 4-year transplant anniversary of Anne Henry)
Elizabeth Hissing (in honor of Erik Hissing)
Henry Holfich
Douglas Holsclaw Jr., M.D. (in honor of Paul Quinton, Ph.D.)
Abigail Huntington (in honor of Rebecca Mueller)
Janet King (in honor of Thaddeus Novak)
Sara Kominsky (in honor of our 20th wedding anniversary)
Judith Lafary
Bonnie Larner
Johanna Libbert
Cathleen Marceau (in honor of Brian Marceau's 30th birthday 9/8/2016)
Jason McDonald
John & Susan McMurry
John Meek (in memory of daughter, Jenny Meek)
Marcia Miller
Loretta Mulatz (in honor of daughter Debra Radler's 54th birthday)
Stephanie Rath (in honor of my 48th birthday on 01/26/2017)
Jeannine Ricci
Scott Rittenbaum
Rob & Ann Robinson (in honor of Carl Wesley Robinson)
Michael Schnitzer
Connie L. Smith
Dorothy Stewart (in memory of Holly R. Stewart, husband & father)
Marilyn Tarr
Robert Tate (in memory of Janice Tate)
Sherry & Bill Thompson (in memory of our daughter, Melissa Thompson-Fleming)
Ted & Susan Tickell
Laura Tillman
Laura Tillman (in memory of Anne Williman)
David Versteeg
Jerome & Laura Watts
David & Pamela Yanero

SILVER

Colleen & Scott Adamson

GOLD

Jonathan Mitz
James R. Yankaskas, M.D.

SUSTAINING PARTNERS

AbbVie Inc.
Cystic Fibrosis Services Inc.

DIAMOND SUSTAINING PARTNER

Nancy Wech (in memory of daughter, Lauren Melissa Kelly, and in honor of son, Scott W. Kelly)

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they found they could prevent the toxins' harmful effect.
http://tinyurl.com/zk52e9n

Can Vitamins Complement Cystic Fibrosis Treatment?

Research has proven that diet modulates gut microbiota in the general population and can bolster immunity and decrease inflammation. Researchers from Australia report that the same may be true for CF patients. The basis of their hypothesis is that CF patients' gut microbiota appears disrupted and may influence pulmonary status. As part of a large study of diet among CF patients, the researchers examined the relationship between micronutrient intake and the gut microbiota. They looked for specific bacterial taxa and micronutrient candidates that might be employed to manipulate the gut microbiota in CF. Their future goal is to identify nutritional therapies that complement CF treatment by modifying gut microbiota composition and/or functions. As CF patients' vitamin C, beta-carotene equivalents, vitamin E, riboflavin and niacin intake increased, their levels of Bacteroides and associated taxa decreased. This is important because the Bacteroides species tends to increase in the gut in patients who experience inflammation. As beta-carotenes and vitamins E and C intakes increased, Firmicutes and Clostridium species also increased; these are species that seem to be affected by elevated oxidative stress

Continued on page 36

How To Move When You Have Cystic Fibrosis

By Devin Wakefield

I have based this how-to on my experience moving from Palo Alto, California, to Seattle, Washington. I, of course, won't have all the answers, while other learned lessons may not even apply to you. However, I do hope you enjoy reading about my experience and perhaps do learn something from my tales.

Some quick background

In June 2016, Microsoft hired me and as part of that I had to relocate to the Redmond, Washington, area. I had lived on the SF Bay Peninsula in California all my life, so this was both a hard and yet obvious answer. My reasons for moving:

- I was living with my parents, and as much as I love them, I wanted to have my own space.
- Rent prices in the SF Bay Area kept me living with my parents.
- I really wanted the new job. My soul was rejecting the commute (it was over an hour each way) plus my job (I did not like the work) plus all my meds (f*@k CF) plus keeping up my living arrangements to my liking (Using clean utensils? What a bother). Get me out of this!
- I'm gay and Seattle also has one of the highest concentrations of LGBT people. My people, I've found you! While San Francisco is nice and has a rich LGBT history and community, so does Seattle–and at a lower housing cost!
- While moving CF centers would suck, I heard good things about the University of Washington Medical Center (UW). I thought it valuable to get the perspective of another set of doctors on my issues with too much mucus everywhere, hemoptysis, bowel obstructions and assaults from alien zombie armies bent on stealing my and others' souls. OK, not that last one, but it may be a nice change of pace.

Where are you going?

We folks with CF don't just move cities, we also move CF centers. As such, I was moving from Stanford to UW. During the interview process with Microsoft, I gave Stanford the heads up. They told me they knew UW to be a fine institution and my social worker gave me the e-mail of the social worker there. Once I got the job, I e-mailed the social worker to get in touch. This turned out to be very helpful, because they got me an appointment before I even had a primary care physician to refer me. This made my transition much simpler. Now I had to find an apartment.

In the apartment hunt, I had some advantages because Microsoft hired me. They have many years experience providing support to the people they hire who have to move hundreds of miles. They gave me some contacts of people who could help me search for an apartment and I greatly prefer that to Craigslist. If you don't have this advantage, do watch out for scams.

DEVIN WAKEFIELD WITH HIS PARENTS' PACKED MINIVAN.

My general tips:

- Get help!!!! I knew a few people in the area, so I contacted them and asked for all their opinions. It's nice to get a general idea of what different areas are like and what areas I might like most.
- Google everything, including maps. I really liked being able to map out where places were before I got there, how far I would be from clinic, work etc. I also had a small list of fun things to do when I got settled.
- Volunteer! I have not done this as much as I want, but I've found it a great way to meet cool folks who care about similar things.

What are you bringing?

Since I lived with my parents, I didn't really have too much to move.

My list:

1. Meds, medication, medicine, medical equipment and more meds
2. Clothes
3. Books
4. Toiletries
5. OH MY GOSH I FORGOT A MED
6. A few personal items
7. Medicines

The lack of furniture to lug 900 miles gave me a big advantage...an advantage I quickly used on handling my medications. Ugh. Gosh, CF, get it together.

Some tips I learned:

- Get a three-month supply of meds before leaving (you can say you're going on a long vacation to get an extra sup-

ply). It's about how long it took me to schedule an appointment with the UW CF team, get referrals and appointments with specialists (sinuses/ENT/otolaryngologist, liver/hepatology, GI/digestive health, endocrinology and dermatology) and find a primary care physician. A three-month supply also gives you one less thing to worry about while moving, unless you forget something important (Ack!! Turn back!).

• I had a portable set of stackable plastic drawers I used to hold my meds, which I could take apart shelf-by-shelf if needed and restack. Very handy organizational tool and if you fill it up with your three-month supply, dense. Something like that may allow for easier unpacking and easier searching.

• Try to get a temporary address ahead of time, in case you have specialty medications you need to ship (or those meds you forgot to get a three-month advance on… oops!) Luckily for me, I had a friend already living up there who had a refrigerator, so I used his.

• Get help when (and I mean when) you need it. It's not worth going it alone.

• Pack a two-week or one-month separate supply of medications (and clothes + toiletries, honestly) as if you're going on vacation. I needed this as the first apartment I thought I had a lock on fell through and it meant I didn't have to go through all my medications and reorganize myself right away. It's organizing extra (fun!) just in case.

Once I got all packed, I hurled it all (gently!) into my parents' minivan and drove off with my father. My dad had left when I moved into my apartment from temporary housing, so I did all the box carrying myself.

When you finish moving:

• Schedule your appointments!!! I felt really weird asking my new clinic for so many referrals, appointments and prescriptions because they didn't know me and why would they do all that for a total stranger? I had to remember that at the very least, it's their job: also my insurance pays them to give care. At best, the clinicians want to help keep patients healthy and they can't do it without you showing up and asking for what you need.

• After moving in your boxes (hopefully all labeled), decide how you'll organize – your Explanation of Benefits, meds, equipment, cleaning supplies and cleaning schedule. Look for local CF groups to get suggestions from other patients and possibly make some friends. Some clinics have a patient group on Facebook and they can help you find good pharmacies, give tips on how best to communicate with the clinic or tell you things to do when you're in-patient.

"After moving in your boxes (hopefully all labeled), decide how you'll organize – your Explanation of Benefits, meds, equipment, cleaning supplies and cleaning schedule."

• When you get furniture at Ikea, try to go early Saturday/Sunday morning. Saturday and Sunday evenings are PACKED! Get off that chair, it's MINE! I saw its low-cost squishy seat first!

Once I secured an apartment, I moved everything in all at once without giving myself much time to rest. Big mistake—I got dehydrated and suffered a bowel obstruction. Whoops! I got my blue medical binder (the fancy kind that you can zip up) out, barfed and luckily a bus line right outside my apartment runs directly to the hospital with my CF clinic (I didn't trust myself to drive very well at that point). I hadn't had an appointment yet, but I didn't know who to call anyway, so in I went.

I am very lucky that I ordered my hospital records from Stanford in both DVD and downloadable formats. I had stored the DVDs in my blue medical binder, forgot about them and then remembered I had them when I opened it up in the E.R. What amazing foresight and brilliance! Wow! (Just kidding.) Although they could access all my Stanford notes and records in Epic, it was still cool to say "here" and hand over a stack of disks when they initially said they didn't have any records about me.

I'm much better now and all acquainted with my CF team—who are VERY cool. My job is going well and other than that GI debacle, I think moving in mostly went fine. I joined a local CrossFit after realizing my exercise regimen got really out of whack during the move. I wanted more structure in my routine and having a buff person decide what exercises to do seemed like a fun idea. I am not sure I would have done it had Microsoft not subsidized me (I LOVE corporate benefits; yay money!), but I am enjoying it so far and hope it is keeping my health up. I am also exercising more than before the move with an adorable trainer. So maybe my gym endorsement is not entirely objective.

I wish all of you who have to move well! Good luck!!! ▲

Devin is 25 and has CF. He moved (Guess where??? Good job! Seattle!) in June 2016 and started living in his apartment on the hot day of August 1st. He still needs a few more chairs that are cushy (and maybe a lamp or two?) for his living room and should really vacuum more often. He is trying, generally, to get it all done reasonably soon. He enjoys hiking through the arboretum near his residence and experienced his first home snow in December!

TILLMAN continued from page 33

seen in CF. Meanwhile, increased dietary potassium decreased Bacteroidales populations. The researchers noted that intake of vitamin and micronutrient supplements at levels considered normal for the general population may not induce gut microbiota in CF patients at levels seen in their study.
http://tinyurl.com/zz4e47q

Researchers uncover a new mechanistic understanding of potential treatment for genetic disorders

A study published by scientists provides insight into the mechanism of action of the drug ataluren, which is showing promise in treating cystic fibrosis. In CF 10 to 15 percent of the single-base pair mutations that cause the disease create a misplaced, premature "stop" codon in the middle of the gene—causing the machinery of the cell to prematurely halt synthesis of the protein, which destroys its ability to function. Ataluren appears to persuade the machinery of the cell to "run" that stop sign and allow a functioning protein to be made. The researchers created numerous premature stop signs, known as nonsense mutations, in test genes in human and yeast cells. They then looked to see what amino acids were inserted when ataluren allowed a skip-over of the premature nonsense mutation. They report that ataluren acts at the ribosome and it allows the insertion of amino acids that are similar to the ones that would have been present in the nonmutated gene. These similar amino acids are carried by what the researchers call near-cognate tRNAs. They also found that the proteins made during ataluren treatment were full-length, and that other aspects of normal protein synthesis were not disrupted.
http://tinyurl.com/zxyfnbn

Colonoscopy, Bone Scans, Diabetes Screening Required for Aging CF Patients

Given that healthcare requirements change as all patients age; cystic fibrosis care teams are now challenged with monitoring evolving health parameters of their CF patients. A growing number of studies show that CF patients are at a high risk for developing adenomatous polyps, which has prompted research into the need for colonoscopy screening. The polyps are benign (non-cancerous) growths that develop on the mucous membrane of the large intestine, but are more likely than other types of colon polyps to become cancerous. The association of cystic fibrosis and adenomatous polyps now requires colonoscopy screening by age 40. Cystic fibrosis patients also exhibit an increased risk for developing secondary cancers. Because breast cancer is a leading cause of death in women, CF patients are urged to get mammograms by the age of 40. Bone loss is another consequence of aging that could be new to older CF patients. Porous bones can lead to diseases such as osteopenia and osteoporosis, which both raise the risk for bone fractures. CF researchers recommend bone densitometry scans every one to five years, depending on the results. Aging CF patients are also seeing more CF-related diabetes (CFRD). Patients should follow the common methods for diabetes assessment, including keeping track of blood glucose, food intake and hemoglobin a1c levels. According to CFRD guidelines, it is important to also screen for complications that can occur with diabetes including retinopathy and hypertension.
http://tinyurl.com/z52cmzq

Exercise Has Central Role in CF Care and Research, Scientist Argues

Substantial health benefits can be gained by cystic fibrosis patients who exercise more, but to accurately assess the impact of such interventions, physicians need to test for exercise capacity and habitual physical activity. Testing provides CF healthcare teams with meaningful information that can be used to tailor interventions, while also providing relevant outcomes in CF clinical trials. Lung function decline in patients who frequently exercise is about 25% lower over time than in those who are inactive.

So, if an intervention is meant to boost patients' exercise capacity, then the amount of moderate or vigorous physical activity that a patient does on a regular basis needs to be assessed and considered. Studies do not necessarily need to focus on physical exercise to benefit from activity testing. For example, studies show that exercise reduces the number of days a patient needs intravenous antibiotics, so to assess the impact of an intervention to reduce antibiotics, researchers need to control patients' physical activity. Additionally, a recent study demonstrated Kalydeco (ivacaftor) improved exercise capacity in CF patients, and that studies are ongoing to explore whether the same holds true for Orkambi (lumacaftor/ivacaftor).
http://tinyurl.com/h5derrw

Exercises Targeting Trunk Muscles May Improve CF Urinary Incontinence

In a recent symposium, scientists discussed the increased importance of physical therapy in addressing CF-associated muscle impairments and urinary incontinence. The role of the physical therapist in managing patients with CF is evolving. At first the physical therapist was restricted to helping patients clear secretions from the lungs. But as patients' life expectancy significantly increases, other body systems also become affected by the disease, such as the neuromuscular and musculoskeletal systems. These secondary impairments caused by CF also need to be addressed so that CF patients have positive quality-of-life experiences. The role of the physical therapist is essential in addressing these problems. Besides clearing secretions and addressing aerobic exercise capacity, physical therapy is needed to help CF patients increase their strength, flexibility and bone density; decrease pain; improve breathing and posture, and help maintain urinary con-

tinence. Researchers said the care team of CF patients should include an active screening strategy for musculoskeletal changes to reduce the impact and prevent, whenever possible, secondary impairments. They emphasized how exercises that aim to mobilize, realign and strengthen the muscles and joints of the trunk (those that move the vertebral column, the muscles that form the thoracic and abdominal walls and those that cover the pelvic outlet) can improve patients' core stability and function. As a result, these exercises may benefit patients by also improving their posture, pain, peripheral muscle weakness, endurance and urinary incontinence. This will ultimately result in better lung function and improved quality of life.
http://tinyurl.com/zdtfmkz

Perceptions Raise Barriers to Palliative Care in CF

A recent study revealed that many patients with CF, as well as parents, are not aware of what palliative care encompasses and how it could be helpful to people with CF and their families. Palliative care is specialized medical care for people with serious diseases, at any stage of the illness. The idea is to provide an extra layer of support, giving symptom relief, and lowering the stress of a serious condition. The goal of such care is to improve the quality of life for both the patient and her or his family. Although CF can be a severe condition that reduces life expectancy among patients, there is no consensus among patients or healthcare professionals about how to incorporate the principles of palliative care into CF management. The research team has developed a standardized education program about palliative care, specific to CF, which it is now testing.
http://tinyurl.com/h2thfpu

CF Women Faced with Lack of Research on Birth Control

There is not enough data to assess the effectiveness of birth control options in women with CF, in whom an unplanned pregnancy may carry substantial risks for both the mom and the fetus. The choice of contraceptives should take into account factors that contribute to the safety and effectiveness of the contraceptive. While most women with CF use oral contraceptives, it's possible this is not the best choice. Many medications boost the activity of liver enzymes that process hormones used in oral contraceptives. If those medications are used together with birth control pills, the concentration of hormones in the blood becomes too low, and there is a possibility that women can get pregnant despite using oral contraceptives. In CF, lumacaftor, included in the combination drug Orkambi, is a strong trigger of these liver enzymes. Women using the drug should not rely on oral contraceptives for birth control. No studies have been performed exploring if and how Orkambi impacts the use of non-oral contraceptives though.

Although most antibiotics (with the exception of rifampin) have no effect on contraception in healthy women, patients with CF are often exposed to high levels of antibiotics and may have a decreased uptake of contraceptive hormones from the gut because of their illness. So far, however, there are no studies that have explored these issues in CF patients. There are also no official guidelines for contraceptive use in women with CF, leaving physicians to take into account disease-specific factors, such as diabetes

Continued on page 38

Former writer, proofreader and Director of USACFA – Anne Williman
May 21, 1953 – September 21, 2016

Anne Williman was a director of USACFA in 2010 and 2011. She wrote articles for *CF Roundtable* and helped with proofreading many issues of the newsletter. She was a prolific writer and wrote more than seven hundred articles, which were published in various magazines. She also wrote two historical fiction books and one non-fiction book, which also were published. She enjoyed being a director, but she found that her health made it difficult to keep up with her duties as a director.

Anne and her husband, Jon, were married for 42 years. Jon was her strong supporter. They had three children and five grandchildren. Anne loved being a wife, mother and grandmother.

Anne had a strong and abiding faith in God and she often wrote of that faith. She said it helped her through the tough times.

One such tough time was in 2016, when she and Jon, along with their son, escaped a very serious house fire. Their dog, who was much loved, did not survive the fire. After the fire, Anne and Jon were staying in an inn, while waiting for their house to be rebuilt. It was very difficult for Anne to be confined to that one room. She was having other health problems and had to be hospitalized. But Anne was not a complainer. She did what she had to and tried to be very compliant. Her last illness proved to be too much for her and she died on September 21, 2016, at the age of 63.

Anne was very happy to have lived so long with CF. She made the most of every day that she had. She is missed by all who knew her.

TILLMAN continued from page 37

and bone health, when selecting suitable birth control options. Studies also show that women with CF prefer to plan their pregnancies well, and that untimely or unintended pregnancies often make them consider an abortion. Despite this, a quarter of all CF pregnancies are unplanned. This can be particularly devastating for a woman with CF, as studies have shown that pregnancy — particularly when a woman has more health issues — can cause increased hospitalizations, a chronic lack of oxygen, fetal growth restriction, preterm delivery, diabetes, heart problems and even death. It is therefore crucial for a woman with CF to be in good health when she becomes pregnant.
http://tinyurl.com/jtvny8o

Severe CF Patients Benefit from Lung Transplant

Patients with very advanced cystic fibrosis (CF) lung disease who get a lung transplant have the same chances of survival as CF patients with more stable disease. The findings suggest that lung transplants are not only appropriate for the sickest patients, but the approach is a sensible resource for CF patients across the board. Although a lung transplant is often the last resort for a CF patient, scientists had raised concerns that a transplant may not benefit the very sickest patients because those with very advanced lung disease may be too sick to survive the transplant.

To investigate the legitimacy of that concern, researchers examined medical records using the lung allocation score to measure severity of disease. To calculate the score, the estimated risk of dying while waiting for a transplant is weighed against the estimated odds of surviving the first year after transplant. The research team found no association between the lung allocation score and worse survival. People with a high score had a one-year survival of 83% and a five-year survival of 65%. For people in the low-score group, the survival chances were 91% and 65% for one and five years, respectively.
http://tinyurl.com/zw7ext3

Key Challenges in CF Vaccine Development Focus of Talk

Developing a vaccine against infections in patients with cystic fibrosis (CF) is challenging, and to succeed researchers need a deeper understanding of the natural history of CF lung infections. Infections in CF can be caused by several microbes, most often bacteria. But studies show that what once was the natural history of these infections has changed. Early antibiotic treatment of Pseudomonas aeruginosa has now made Staphylococcus aureus the most commonly isolated bacteria in CF patients. Infections with mucoid P. aeruginosa, however, remain the major initiator and driver of lung function decline in CF, and vaccination against this microbe is still a research priority.

When developing vaccines, researchers aim to identify epitopes — parts of surface proteins on bacteria — that if targeted are most likely to give results. But this approach is problematic, since bacteria tend to evolve rapidly to evade immune responses, changing their epitopes along the way. Researchers have also experimented with so-called live attenuated vaccines. Such vaccines are composed of versions of a microbe engineered to induce an immune response but not able to cause disease. This may not be the best approach for CF patients, however; scientists are concerned that an active vaccine that triggers T-cell immune responses can increase lung inflammation in patients. New techniques to assess the composition of bacterial species also reveal that the bacterial communities in CF airways are quite complex. As of now, researchers do not know how some of the newly identified species contribute to disease, or how the removal of one species would affect the composition of the remaining microbes. In addition to classical vaccine strategies, researchers have considered treating patients with antibodies, an approach often called passive vaccination. Unlike traditional vaccines, antibodies can also be used during an infection.
http://tinyurl.com/hlo6uhk

Life-threatening superbug spreads globally in cystic fibrosis patients

A multidrug-resistant superbug infection that can cause life-threatening illness in people with CF has spread globally and is becoming increasingly virulent. The researchers said the bug, a species of multidrug-resistant bacteria called Mycobacterium abscessus (M. abscessus), can cause severe pneumonia and is particularly dangerous for patients with CF and other lung diseases. The bug initially seems to have entered the patient population from the environment, but it has recently evolved to become capable of jumping from patient to patient, getting more virulent as it does so. They found that the majority of patients had picked up transmissible forms of M. abscessus that had spread globally. Further analysis suggested the infection may be transmitted within hospitals via contaminated surfaces and through the air, the researchers said—presenting a serious challenge to infection control practices in hospitals. Because the superbug has already become resistant to many antibiotics, it is also extremely difficult to treat successfully. Patients infected with it need 18 months or more of treatment with a combination of powerful antibiotics, and fewer than one in three cases is cured.
http://tinyurl.com/z8f2xc4

Scientists in Ireland developing new lung disease treatment

Scientists in Ireland are developing a new treatment for lung diseases, including CF and asthma. The first of its kind dry powder inhaler will deliver an innovative drug into the lungs in a way that is hoped will effectively break up thick sticky mucus that can cause lung infections.
http://tinyurl.com/gwn9ryr

TGV-inhalonix To Report New Data on Breakthrough Drug for Cystic Fibrosis

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at the Largest Conference of the Year

Mul-1867 has already shown promising efficacy in human studies outside the U.S. The U.S. Food and Drug Administration earlier this year granted Mul-1867 Orphan Drug Designation based on early indications of its potential for fighting lung infections in CF patients. The special status is intended to provide incentives for the development of drugs and biologics to treat rare diseases. Mul-1867 is the first drug candidate under development that holds promise against multiple strains of bacteria, as well as those infections caused by a mix of bacteria and fungi.
http://tinyurl.com/zjnh7d6

OrPro Therapeutics and Collaborators Present Key Theradux™ Data

Theradux was found to be at least twice as effective in increasing mucus transport than Pulmozyme® (human DNase, dornase alfa), a standard-of-care mucolytic whose target is viscous DNA released from cells and bacteria as a consequence of chronic infection and inflammation. While not a cure, such an approach may, like CFTR modifier drugs Kalydeco® (ivacaftor) and Orkambi® (lumacaftor and ivacaftor), act to alter the course of disease and mitigate onset

Continued on page 40

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- 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.
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- 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.
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- 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.**
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TILLMAN continued from page 39

of the serious late-stage lung symptoms that are the primary contributors to CF morbidity and mortality.

Theradux represents a new class of inhaled, non-systemic therapeutics that target underlying structural changes in CF mucus via a unique pH-independent binding mechanism that both prolongs activity and sequesters the drug in the mucus layer. Theradux relaxes excess protein linkages without disrupting the polymeric mucus gel structure essential for normal transport and clearance. A biologic drug derived from a native airway enzyme (thioredoxin), Theradux is hundreds of times more active than small-molecule mucolytic agents, with far greater selectivity and specificity for mucus protein disulfide bonds.
http://tinyurl.com/jkrrtyc

FYI

Cancer risk among lung transplant recipients with cystic fibrosis. Fink AK, Yanik EL, Marshall BC, Wilschanski M, Lynch CF, Copeland G, Safaeian M, Engels EA. J Cyst Fibros. 2016 Aug 15.[Epub ahead of print]

Previous studies demonstrated increased digestive tract cancers among individuals with CF, particularly among lung transplant recipients. Standardized incidence ratios (SIRs) compared cancer incidence to the general population and competing risk methods were used for the cumulative incidence of colorectal cancer. The results indicate that CF recipients have increased risk for colorectal cancer, suggesting a need for enhanced screening.
http://tinyurl.com/greynuc

SINUSITIS

Paranasal Sinus Size Is Decreased In CFTR Heterozygotes With Chronic Rhinosinusitis. Calton JB, Koripella PC, Willis AL,Le CH, Chiu AG, Chang EH. Int Forum Allergy Rhinol. 2016 Nov 17.

Cystic fibrosis (CF) heterozygotes with a single mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene are at significantly higher risk to develop chronic rhinosinusitis (CRS). However, the reasons why remain unknown. The hypothesis that CFTR heterozygotes would have smaller sinus volumes than healthy controls was tested. To exclude sinus disease as a confounding factor paranasal sinus volume in those with CRS, but without known CFTR mutations was also assessed. CFTR heterozygotes with CRS have significantly smaller frontal and maxillary sinus size compared to those without mutations, irrespective of disease state. This sinus hypoplasia may contribute to impaired mucus clearance and chronic sinus disease development.
http://tinyurl.com/h9pd87o ▲

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