CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
SUMMER 2014

That's My Plant-based Diet And I'm Sticking With It!

By Tom Martin

TOM MARTIN BIKING ON THE SANTA CRUZ BLUFFS.

I wrote an article titled "Go Vegan And Nobody Gets Hurt" that was published in the Autumn 2013 issue of *CF Roundtable*. I feel like I need to follow up and talk about my progress since then.

First a little about myself: I grew up in Santa Cruz, CA, and currently live in Aptos. I'm an avid mountain biker, and I work from home as a full-time Mechanical Designer Drafter. I have my own business at Martin Design and Drafting Consulting.

I'm excited to say, on the one-year anniversary, my plant-based diet has been very good to me for the following reasons. The most important is that I have no constipation and no bowel obstructions; this is a big deal for me. Prior to my vegan diet I was bloated after a meal and chronically constipated, hence the pain after eating. Secondly, I have had no weight loss; I've maintained my BMI of 24.5 and I'm staying steady. I found that the trick is to load up on potatoes, bread, pasta, and beans/lentils and eat lots of avocados for good fats and calories. I really like to sprinkle salt and pepper onto a fresh cut avocado; it's a delicious way to keep the body weight up.

Complex carbs are good for long-term energy. I've heard the body needs at least 1,000 calories minimum to keep the body's organs func-

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INSIDE THIS ISSUE

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EDITOR'S NOTES

Summer is here and is treating us well. The weather has been a little hotter than I like, but we have had occasional days of rain to keep everything green. How I love that green.

I am sad to report that **Laura Tillman** is no longer a Director of USACFA. She decided not to run for another two-year term. She also resigned as President, effective June 8, 2014. We all will miss her leadership. She still will compile the "Information from the Internet" so we still will be hearing from her. We thank her for all her years of service and wish her well in her retirement from USACFA. **Jeanie Hanley** will finish out Laura's term as President. Congratulations, Jeanie.

We are happy to welcome a new Director. Her name is **Karen Vega** and she sounds as if she is full of energy and ideas. We look forward to working with her. You may read about her on page 14.

I hope you have read the follow-up article **Tom Martin** wrote about his experiment with a vegan diet. It sounds as if it's working for him.

Beth Sufian, in "Ask the Attorney," answers questions about Medicare and Medicaid that have been submitted by our readers.

Our Focus topic is: Dealing With Conditions That Are Part Of CF. **Campbell Bryan** discusses the danger of injuries that can be caused by the exercise that is so important to our well-being. In her article, **Andrea Eisenman** talks of her use of an insulin pump to help manage her diabetes. In "Coughing With A Smile," **Jennifer Hale** tells of her efforts to maintain her blood sugar and working with decreasing lung function. **Julie Desch** uses her "Wellness" column to discuss some of the less talked about aspects of having CF. (Definitely applies to all of us.) I continue the topic with reference to several problems that many of us share.

"In the Spotlight," by Andrea Eisenman and Jeanie Hanley, features **Emily Schaller**. She does so much that just reading about all of it makes me tired.

Isabel Stenzel Byrnes writes of discipline and how it relates to our CF in "Spirit Medicine." **Paul Feld** reviews a book that was written by the father of a man who had CF. On page 18, be sure to check out the notice about a new Website for CF mothers.

USACFA sends our congratulations to **Dr. Jerry Nick**. Read all about it on page 27.

We still welcome your donations to our Pay It Forward campaign. Your continued support overwhelms us. Thank you.

Please check out "Looking Ahead" on page 3 to see if there is a topic that piques your interest. In our next issue, we will feature articles that are written by adults who have CF, but we will accept articles by family or friends of adults who have succumbed to CF. We cannot guarantee that all articles will be published.

Until next time…stay healthy and happy.

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

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

Paul Feld
Florissant, MO
57 on May 9, 2014

David Lewis
Elk Grove, CA
46 on May 31, 2014

Debra Radler
Roselle, IL
52 on May 31, 2014

Anne Williman
Middletown, OH
61 On May 21, 2014

Wedding

Amy & Robert Branham
Fairmont, WV
7 years on June 30, 2014

Paul & Kristi Feld
Florissant, MO
23 years on June 1, 2014

Debra Radler & Adrian Gulinski
Roselle, IL
7 years on June 8, 2014

Transplant

Andrea Eisenman, 49
New York, NY
Bilateral lungs
14 years on April 25, 2014

NEW BEGINNINGS

Amy Branham, 38
Fairmont, WV
January 13, 2014
Cancerous thyroid, surgically removed
February 17, 2014
First radiation treatment
Now is cancer free.

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**

Summer (Current) 2014: Dealing With Conditions That Are Part Of CF.

Autumn (November) 2014: Dealing With The Death Of A Loved One With CF. (Submissions due September 15, 2014.) Have you had a spouse, sibling, child, parent or friend die from CF? How did that death affect you? How did you cope with the death? Do you have any suggestions to make it easier for others to handle?

Winter (February) 2015: Ways To Become A Parent When You Have CF. (Submissions due December 15, 2014.) Are you a parent who has CF? What method did you use to become a parent? Do you have suggestions for others who want to become parents?

Spring (May) 2015: Transitions – Many Types of Changes. (Submissions due March 15, 2015.)

ASK THE ATTORNEY

Answers To Readers' Questions

By Beth Sufian, JD

In the past three months readers have asked many questions about Medicare. As more adults with CF work, more adults become eligible for Social Security Disability benefits if they become unable to work or stop working in order to spend more time with daily CF medical care. Medicare typically provides good coverage for CF medical needs and other medical needs. However, there are some quirks in terms of which part of Medicare covers which CF medication. See below for more information. In addition, the first question answered in this column concerns the Social Security proposed new medical criteria.

CF Roundtable *readers can contact the CF Legal Information Hotline at 1-800-622-0385 or CFLegal@sufianpassamano.com if there are additional questions related to Medicare, accessing insurance coverage through the federal or a state Healthcare Exchange, questions about Social Security benefits or questions about rights in education or employment.*

1. When will the Social Security Administration (SSA) issue new cystic fibrosis (CF) medical criteria to be used when determining eligibility for Social Security benefits?

In April 2013, the Social Security Administration held a conference call for the CF community and estimated that new CF eligibility medical criteria would be published in July of 2014. However, Social Security representatives now indicate that the CF medical criteria are expected to be published in November or December 2014. No indication of which proposed changes will actually be made was noted by the Social Security representatives.

Individuals who are concerned they might not meet the new SSA medical criteria should remember that the medical criteria SSA uses to determine if a person is eligible for benefits are the medical criteria in force at the time a final decision is made on the application. If a person's application for SSA benefits is pending at the time new medical criteria go into effect, then the application will be assessed using the new medical criteria. It typically takes three to six months before an application is approved. Therefore, if a person applies for SSA benefits in August of 2014 the application may not be decided until January 2015. If new medical criteria are published on November 1, 2014, the criteria will go into effect 30 days later on December 1, 2014. An application filed in September 2014 that is decided in January 2015 will be decided based on the new medical criteria.

BETH SUFIAN

2. Which states have expanded Medicaid under the Affordable Care Act (also referred to as Obamacare by many)?

The following states have expanded Medicaid eligibility to adults who are not on SSI and meet certain low income criteria: Arizona, Arkansas, California, Colorado, Connecticut, Delaware, D.C., Hawaii, Illinois, Iowa, Kentucky, Maryland, Massachusetts, Michigan, Minnesota, Nevada, New Jersey, New Hampshire, New Mexico, New York, North Dakota, Ohio, Oregon, Rhode Island, Vermont, Washington and West Virginia.

There are four states that are still actively considering expanding Medicaid: Indiana, Pennsylvania, Utah and Virginia. All other states have said they will not expand Medicaid at this time. If a person with CF lives in a state that has expanded Medicaid and is eligible for Medicaid benefits but then moves to another state where Medicaid is offered only to individuals who receive SSI benefits, then the person with CF will lose his/her Medicaid benefits unless he/she is also receiving SSI benefits.

3. If Medicare does not pay for my transplant surgery because I have private insurance at the time of the transplant, will Medicare pay for my transplant medications if in the future I lose my private insurance coverage and have only Medicare?

Yes. If Medicare did not pay for the transplant then Medicare Part D will provide coverage for transplant drugs in the event Medicare becomes the primary insurance coverage or the only insurance coverage a person has to pay for medication. If Medicare pays for the transplant, then Medicare Part B

will pay for the transplant medications. Medicare Part B typically has a 20% co-pay for medications.

Medicare Part D has a more complex payment structure. Medicare Part D pays for medication up to a certain amount of money and then a person is in something called the "donut hole." While in the donut hole, the person pays 100% of his/her medication costs. The Affordable Care Act is reducing the donut hole each year until, in 2020, the donut hole will only be $250. Once a person pays his/her out-of-pocket share and is out of the donut hole, then the person is in Medicare Part D catastrophic coverage and most medications have a co-pay of between $5 and $20 a month. Some medications may have a co-pay of 5% of the cost of the medication. It is possible to obtain co-pay assistance from a manufacturer that will pay for the cost of drugs needed while in the donut hole or if there is a 5% co-pay once a person is in Part D catastrophic coverage.

> *"There is no Medicare coverage for people with CF for home IV supplies or home nursing while on home IVs."*

4. Does Medicare cover home IVs for people with CF?

Medicare Part D plans will provide coverage for IV medication. There is no Medicare coverage for people with CF for home IV supplies or home nursing while on home IVs. In order to have home IV supplies or home nursing covered, the home IV medication used has to be covered by Medicare Part B. Part B covers home IVs for only approximately eight diseases. CF is not one of the listed diseases.

Nothing in this column is meant to be legal advice about your specific situation. If you have additional questions, please contact the CF Legal Information Hotline at 1-800-622-0385. The Hotline provides free and confidential legal information to people with CF, their CF Care Center teams and their families. The Hotline is sponsored through a grant from the CF Foundation. Callers speak to an attorney employed by the Hotline. The Hotline is the only service that provides legal information from attorneys who focus their practice on the rights of people with CF. The Hotline can also be reached by e-mail at CFLegal@sufianpassamano.com. The Hotline has handled more than 34,000 calls from people with CF, their family members and their healthcare providers. ▲

Beth is 47 and has CF. She is an attorney who specializes in disability law and she is a Director of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues.

Information For People Who Travel On Airlines

In December of 2011, the Transportation Safety Administration (TSA) introduced a toll-free helpline that answers questions for fliers with disabilities and medical conditions. Disabled travelers may call ahead about screening policies, procedures, and what to expect at the security checkpoint. The purpose of the new helpline is to inform passengers with disabilities about certain policies before they fly so they may properly prepare for travel. The helpline may be helpful for people with cystic fibrosis who must travel with durable medical equipment, portable breathing machines, needles, liquid solutions (inhalant medications, insulin, etc.) and other medical equipment.

Travelers are encouraged to call at least 72 hours prior to a flight. People with CF and other medical conditions may call the "TSA Cares" toll-free number at: 1-855-787-2227. The helpline is available Monday through Friday, 9am-9pm (Eastern Time) and is closed on all Federal holidays.

Individuals also may find information on traveling with special medical needs on the TSA Website at: http://www.tsa.gov/travelers/airtravel/disabilityandmedicalneeds/tsa_cares.shtm.

SPIRIT MEDICINE

Gaman With Your 'Ness: The Spirit of Pushing

By Isabel Stenzel Byrnes

"*The brick walls are there for a reason. They're not there to keep us out, they're there to let us prove how badly we want things.*"
– Randy Pausch, 2008

In this article, I'd like to muse about the spirituality of perseverance. By perseverance, I mean the acts of exerting effort, having grit, discipline, endurance, or pushing oneself – through goals, achievements and day-to-day "brick walls" that we all face. We in the CF community, more than any others, are shining examples of perseverance! Our day-to-day lives are filled with opportunities to practice perseverance. In fact, writing this article is one of those. I don't always feel like it, but as the deadline approaches, I feel pressured, and I know I'll enjoy the process and the final product. So I force myself to write.

As a half-German, half-Japanese woman, I come from cultures where discipline and hard work were deeply engrained into me as a child. As I got older, my twin, Ana, and I developed our own twin language. One of our most common phrases was "*gaman with your 'ness*." In Japanese, the word *gaman* means endure through discomfort. The "-ness" referred to the suffix attached to an adjective to make it a noun; it denotes a quality, condition or state; or, for our purposes, a sense of "being" or "essence," like "happiness" or "kindness." So we would often tell each other to just use our whole essence to hang tight, to endure with whatever it was we were facing.

Besides my ethnic background, my CF culture became more influential in the development of my disciplined character. Like most people with CF, I was taught at a very early age that life consisted of doing uncomfortable things I didn't want to do (i.e., treatments), but had to do, and that not doing those things would result in a bad outcome. Being extremely disciplined with treatments was one way to reduce the chances of a threatened lifespan.

There was nothing passive about treatments. I remember during my hours of chest physical therapy, my mother would say, "Don't just lie there, make an effort to cough!" She would add, "If I'm making an effort, you must make an effort!" Indeed, CF required everyone in the family to be disciplined. And so, every day of my life, I learned that pushing myself *hard* to cough, to stay up late or get up early to do treatments, to eat when I didn't feel like it and to exercise were normal.

ISABEL STENZEL BYRNES

Discipline in the CF lifestyle helped to reduce my death anxiety. Exerting my maximum effort in my healthcare meant I was doing everything I could do to control my fate; the rest I surrendered. This gave me hope. There is a scripture that reads, "*More than that, we rejoice in our sufferings, knowing that suffering produces endurance, and endurance produces character and character produces hope.*" (Romans 5:3-4) This quotation rings so true in the lives of so many of my CF friends.

The discipline required for the CF lifestyle crosses over to other aspects of life. I love reading stories of CF peers who get up super early to do treatments before work or school, or who are high achievers in school, work and life demands. I read about those who cultivate the discipline to train for a marathon or triathlon. I am deeply inspired by the collective effort that our community exerts to live the best life we can all live. We are a feisty bunch!

Having athletic mentors helps me to reach my own fitness goals. I am now training for swimming competitions at the Transplant Games of America, which will take place in July 2014. Each time I practice a sprint race, I exert 90-100% of effort, and my muscles burn, my heart pounds and I finish the length with a huge gasp followed by heavy panting. It doesn't feel good. Pushing myself hurts.

Exerting effort often comes with discomfort in the moment. The reward comes in the future – in the

form of greater fitness (and for some, better looks), longer lifespan and better quality of life. Exerting effort in school or work often leads to richer and better opportunities in the future. And thankfully, pushing ourselves has potential to increase self-esteem by helping us feel accomplished, determined, successful, even defiant. Pushing ourselves helps us prove to others that we "got what it takes."

Discipline comes in many forms. Last month, I accomplished the hardest task of my life. I gave a short TEDx talk in front of 650 people at Stanford University, on an emotionally-loaded topic, "The Art of Saying Goodbye." I gave this talk eight months after Ana died, and in my grief I wasn't quite ready to present to the public. However, I had to harness my lifetime of discipline to push myself – emotionally and spiritually – through my own loss-confusion, to give this honorable speech. Afterwards, I felt immense relief that it was over, and I felt proud of myself for "gaman-ing with my 'ness."

Perseverance and discipline are seen as virtues in many faiths. We often find ourselves praised by society for hard work and steadfastness. This external validation motivates us to remain focused. But when is endurance and discipline not healthy? As I get older, I realize that intense athletic pushing or masochistic discipline overwhelms me. Long-term pushing and discipline can be a form of stress – which can slowly eat away at our health and balance. Discipline that is fueled by a reactive and zealous urgency – like running away from fear or other difficult emotion, or desperately trying to be held in positive regard by another – can be toxic. This type of discipline contaminates the present moment, infusing it with anxiety.

Discipline that leads to physical harm is not good (like having a big lung bleed at work, and then just continuing to work afterwards – yep, that was me). Also, pushing oneself with the *expectation* of a reward may lead to disappointment. Sometimes great effort fails to produce the wished-for outcome – we've all heard of someone fighting CF so hard, but dying before donor lungs become available. In these cases, effort is for effort's sake; to practice perseverance is the lesson.

Finally, pushing oneself becomes harmful when *not pushing* leads to self-loathing and self-judgment. Then we start "should-ing all over ourselves." Every human being has his or her own threshold for "enough," and for a limit to tolerating discomfort. I know I have to forgive myself when I'm just too tired or don't feel like working out. I have to listen to my instinct that something else is needed instead. Giving into fatigue, or laziness, is not a flaw. I firmly believe we cannot exert 100% of the time. There is a time for pushing and a time for rest. We need to allow ourselves to let go, to take a break, to relax sometimes. (Even if it's contrary to our CF nature!) This restoration and renewal will offer centering, and even recharging for our next big effort.

Pushing ourselves can be seen as a spiritual practice. Spiritual discipline usually pertains to spiritual practice in the form of prayer, study of scripture, ritual or meditation. But effort of any kind – to aim for a promotion, to achieve athletic prowess, to get through this exacerbation – is an invitation for us to reach a deeper level of spiritual awareness. The deepest, truest discipline has its roots in the ancient wisdom of the Hebrew prophet Zechariah: "*Not by might, nor by power, but by… spirit.*" It takes a rewiring of our thinking, but harnessing and cultivating spiritual stamina may be exactly what we need to help us push through physical, intellectual or emotional brick walls.

And yet, truthfully, sometimes it is very difficult to be disciplined. It requires strong mental focus and positivity. Having CF can take a toll on our bodies but also on our minds, emotions and spirits. When we are feeling really sick, depressed or exhausted, we might feel too depleted to push. Sometimes it could help to look outside of oneself and see one's effort for a greater purpose or cause. There are spiritual quotes from every faith that might inspire us. For example, the Quran says, "*Verily, with every difficulty there is relief. Verily, with every difficulty there is relief.*" (Chapter 94, Verse 5-6). Sometimes we just need reassurance.

Seeing yourself as a spiritual instrument can be helpful in infusing inner discipline. Some people with CF run marathons to show the Universe that people with CF can achieve such monumental feats. I also try to see my own efforts in whatever goal I'm facing as a way to honor those who didn't have the chance to be here – like my organ donor and so many CF peers. And, if some can't push themselves for themselves, or for their family, some people might be able to push themselves for God. The Universe welcomes all contributions to a greater collective spiritual effort.

And finally, if we can't push further, it's okay to surrender to the moment, to be okay just as is and to divert efforts to a spiritual focus. In spirit, perseverance means we did our best. God and the Universe will still love and respect us for who we are, not what we are able to do. It takes true discipline to be at peace with who and what we can be. ▲

Isabel Stenzel Byrnes is 42 years old and 10 years post-lung transplant. She lives in Redwood City, CA. Just to share, her TEDx talk can be seen at https://www.youtube.com/watch?v=Dkffpibi-Dc.

SPEEDING PAST 50...

CF Can Be A Full-time Job

By Kathy Russell

Summer is here and I'm ready for it. Recently, I had cataracts removed from both eyes and new lenses implanted. Wow! I love being able to see everything. The trees have so many leaves! I really hadn't seen them for many years. For the first time in my life, I have 20/20 vision. It is amazing. Now, instead of taking my glasses off to read, I have to put on reading glasses. How cool is that? Life is good.

I always feel better in the summertime. I have allergies, but I find that taking loratadine helps me manage the symptoms and the warmer weather feels so nice. I know that summer means hot and humid to many of our readers. For you, I have great sympathy. Where I live, in northern Oregon, our temperatures are in the moderate range and I love it. And even more important, it cools off at night.

When the weather is nice, I find it difficult to do my chores. Writing this column, for instance, becomes a chore. I would rather be out enjoying the lovely weather. But, to be honest, I wouldn't be sitting outside anyway. I am on continuous supplemental oxygen and find it easier to stay inside. I know that I don't have to stay inside just because I use oxygen, but I find that I do.

I suppose that I use CF and oxygen as excuses to do less than I might otherwise. I have less energy than I used to and am not as interested in doing things as I once was. After taking care of my CF needs, I am just tired. It is as if CF is my job.

There are times that having CF can be a full-time job. What with all of the meds and treatments, we spend a lot of hours dealing with CF. If we have to deal with insurance coverage, we may spend hours on the telephone trying to get wrinkles ironed out. I am fortunate to have a wonderful husband, Paul, who is my advocate and handles all of those annoying calls.

The Focus topic of this issue is: Dealing With Conditions That Are Part Of CF. Many of us have other health problems that are directly related to CF. One of my main "conditions" is the use of supplemental oxygen (O_2). I have a large O_2 concentrator at home and a smaller O_2 concentrator that I take with me when I leave home. We had to go through so many hoops to get Medicare to provide what I wanted.

We had jumped through many hoops several years before, when I first got O_2, because my private insurance thought I didn't need to have a source of oxygen that would allow me to leave home. I told them that I didn't think there were any extension cords that were long enough to let me get to the doctor's office or to my grocery store. They relented and I got my portable concentrator. Thank goodness for Paul! So, we were prepared for the fight with Medicare. It took time and energy to accomplish, but the result was worth the effort.

> *"Another problem with our coughs is that we can lose consciousness. After several hard coughs, I see black with multi-colored stars."*

KATHY RUSSELL

One of the most common issues of CF is sinus disease. I have had way too many years of dealing with rotten sinuses. The terrible pain and foul odors in my head were most unpleasant. You'll notice that I said, "…were most unpleasant." That is because those days are in the past.

Since my last sinus surgery, nearly 20 years ago, I have had no further troubles with my sinuses. After I was healed up from that surgery, I started irrigating my sinuses every day, without fail. I use a sterile normal saline solution and a little bit of sterile glacial acetic acid to do the irrigations. That has been enough to keep from getting infections in my sinuses. I don't miss the headaches and terrible pressure that I used to feel. Having almost normal sinuses is great!

Another common problem that is a part of CF is gastro-esophageal reflux disease or GERD. Those of us who

have dealt with GERD know how debilitating it can be. Having acid splashing up into the back of one's mouth, and spraying into the lungs on its way, can cause all sorts of erosions and even pneumonia. That is true for anyone, but when one has CF it is even more of a problem.

I had bad scarring in my esophagus and had suffered with ulcers. After getting started on Nexium, my reflux eased up. I don't miss the pains that used to wake me from a sound sleep, as my intestines would twist. It is a few years since I have experienced that pain and I am delighted to do without it.

Other gastro-intestinal problems include diarrhea, constipation, steatorrhea, bloating, hemorrhoids, rectal prolapse and…you get the idea. All of us can experience any or all of them at one time or another. Some of the things that ease these problems include pancreatic enzymes, laxatives (such as Miralax), probiotics (including natural yogurt), anti-gas preparations (such as simethicone), medicated suppositories, surgery and so on.

Then there are fun things such as cranky gall bladders that eventually must be removed. (Been there, done that.) When I had part of my colon removed, the surgeon removed my gall bladder at the same time. I don't miss the pain under my right ribs from having an inflamed gall bladder.

I have taken enzymes, at times, but I haven't needed them in quite a few years. Evidently my pancreas works when it wants to. Fortunately for me, it has been working for several years now.

So we've talked of sinuses, GERD and other gastro-intestinal issues. Now we'll mention CF-related arthritis. That has caused me so much distress. My spine is twisted and curved. My hands and feet ache almost constantly. I experience pain in my hips, knees and ankles. I find that keeping my bones warm and not flexing or over-extending my joints help to avoid pain.

When I was young I used to walk a lot, exercise plenty and often overdo activities. I paid for that with terrible leg aches and occasional cramping of muscles. Now I use better judgment and have fewer bouts of cramping and pain.

One thing that I have done to be nice to my skeleton is getting custom orthotic devices for my shoes. Having the proper support for my feet (and for my high insteps) has given me relief from some pain. These also have the added benefit of helping me to stand straighter, walk easier and not twist my ankles. Every little bit helps.

Another item that has been of assistance in easing my pain is a good mattress. For the past dozen-or-so years, we have slept on a Tempur Pedic mattress. It is expensive to purchase, but the quality is worth the price. It doesn't cause any sore spots on my body, and it gives excellent support. My back gives me much less trouble than when I sleep on any other kind of mattress. When I wake up, I can get out of bed and walk with little difficulty. That, alone, is worth the price.

A problem that plagued me when I was young was excessive sweat. (I know, "horses sweat, men perspire and women glow," but I sweat!) I wore out shoes so fast because of my sweaty feet. While I was working as a nurse, I bought new work shoes every six weeks. My sweat would cause the leather to crumble. First it would cause ugly staining, then the leather just crystalized and broke apart.

When I started wearing European-style clogs, I had less trouble with sweat damage. These shoes were open at the back and my feet didn't get so hot. It really helped. I still wear shoes that let my feet breathe. If I wear closed shoes, it is as if my feet are sitting in warm salt water and they get all wrinkled.

I haven't mentioned anything to do with our coughing. There are problems that are caused by our coughs. Just recently, I experienced a prolonged bout of coughing that felt as if I tore a muscle in my back. It hurt so much that I told Paul that I had to lie down. I did and I fell asleep for two hours. When I awoke, I felt okay. Sleep is a wonderful restorative.

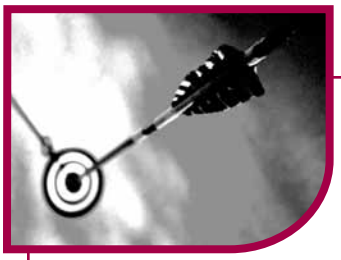
Another problem with our coughs is that we can lose consciousness. After several hard coughs, I see black with multi-colored stars. I know that I am about to pass out, so I concentrate on getting some air. Since I have oxygen blowing into my nose all the time, it easier for me than for some others. I take a few breaths and am ready to face the world again. Another problem averted.

I won't even try to figure out a treatment for the occasional loss of continence that can be caused by our coughing. I know that all of us have had to deal with having at least a few drops of urine escape with hard coughs. No matter how many Kegel exercises we do, we will have some incontinence. It goes with the coughs.

I know that I have forgotten some of the troubles that CF has caused for me. That is fine. If I can't remember them, they must not be bothering me now. All of our problems can be managed fairly well. You and your healthcare team have to work on finding the right treatments for each problem. Also, we are fortunate enough to outgrow some of our problems. I hope that you can outgrow all of the ones that give you the most discomfort.

Now that this chore is done, I'm going to look at everything that I can and really enjoy my new ability to see. Life is good and I love being alive. ▲

Kathy is 70 and has CF. She and her husband, Paul, live in Gresham, OR. You may contact her at: krussell@usacfa.org.



FOCUS TOPIC

DEALING WITH CONDITIONS THAT ARE PART OF CF

Exercising Into Injury

By Campbell Bryan

I am a 30-year-old female with CF. I was diagnosed at 17 years of age and have mild symptoms. I am a very active person, who is always on the go and loves exercise. One could even say that my love for exercise could have fortunately (or unfortunately) been related to my late diagnosis.

I want to discuss the fact that exercise is medicine for an aging population of adult CF patients who demand a lot of their bodies daily. Increasing physical demands on our bodies causes wear and tear and increases chances for injury, thus my constant challenge of avoiding injury while pushing my body to the limits (and the limits are starting to be more limiting). I feel this topic does not get enough attention; and I hope raising it will help others focus on sharing information regarding the potential for injuries caused by exercise, ways to avoid injury, best practices for lung-clearing exercises with least demand on the body, and access to those professionals who may help aging adults better manage physical injuries.

I have always been a runner,

CAMPBELL BRYAN

which has kept my lung functions above 80. A year ago I suffered peroneal tendonitis in my ankle and had to stop running. I didn't have easy access to a pool, so my lung function dropped 10%, super-fast! It was very scary and hard to think about how long it would be before my ankle would be ready for all the miles I needed to get in. Mentally and physically, this was a huge challenge. I was desperately racing toward a solution.

I worked with a physical therapist and tried my hardest to follow the routines. At least I was able to walk without pain during the healing time. I got a treadmill in my house and now I do 30 minutes morning and night on the treadmill (incline of 10, speed of 3.1) while doing my vest. Wearing the vest while walking briskly on an inclined treadmill simulates a run and really provides the boost I need. My lung function is back up to above 80, and my ankle is ready for a few runs.

The thought I want to spark in others is that I believe my injury easily could have been prevented. When we make sure to push CF children and teens to exercise, do we also push them to stretch and educate them on preventing injuries as much as possible?

I thought I was a good athlete and ran five miles a day through college, not realizing that each year my muscles were changing due to my lack of stretching. Now I am working hard to re-train my muscles, especially since my lungs now are harder to maintain than they were in my early 20s. I hope we can make sure other athletes out there get the information they need to prevent injuries or to, at least, heal faster. ▲

Campbell is 30 and has CF. You may contact her at: cbryan83@gmail.com.

Pay It Forward

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

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

Pauline Dinello
Bryan Dudman
Tonya Hamilton
Richard Harris
Mel & Marion Baldwin Hayes
Jennifer Staashelm

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YOU CANNOT FAIL

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

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

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

Trying New Things To Control Old CF-Related Problems

As many people with cystic fibrosis (CF), I developed CF-related diabetes (CFRD). But it appeared only after my bilateral lung transplant in 2000. So, at 35, at the time of my surgery, I was probably borderline diabetic. It was probably the high doses of prednisone that were administered to me first by IV and then in pill form post-transplant that sent my pancreas packing.

Being diagnosed with CFRD a few days after transplant, as I was learning about my new medications for my new lungs, was all so overwhelming. I had no idea about counting carbs or giving myself insulin injections, let alone taking immuno-suppressants. It was a rocky first year with my new-found diabetes. If it had not been for the late Pammie Post, a former director of USACFA and all around wonderful person, who gave me an instructional manual for diabetes and talked to me at times for hours, I do not think I would have grasped it. Pammie was a maven who had had diabetes before her bilateral lung transplant and she was in her 50s. She knew it all well and was always ready to assist others.

After talking to Pammie and being taught by a diabetes educator, I eventually started to get it. My numbers fluctuated from 50s to 200+. But I learned from Pammie what to eat or drink for a glucose low and foods that were diabetic friendly for weight gain due to the malabsorption from CF. Most diabetic educators are not versed in the fact that we cannot be on a low-carbohydrate diet and need to gain weight, unlike many other type-1 or type-2 diabetics.

After a few years of transplant medications and giving myself insulin injections, I felt I had it under control. My A1c was okay, usually around 6.5 or in that vicinity, which I felt good about. But every time I saw my endocrinologist, she would suggest I try an insulin pump. For some reason, that terrified me. I used to have a port-a-cath and flushing it once a month was always stressful. And having something in or on my body at all times was not something I looked forward to.

ANDREA EISENMAN HOLDING HER INSULIN PUMP

> "I just saw my endocrinologist and she was thrilled that: a) I was enjoying the new pump, and b) my control was good."

But, since she is my doctor and I respected her, I started to ask my friends with CF who manage their diabetes on a pump system about their experiences. Mostly, they were positive. Also, many friends without CF who I consulted acted like I was nuts NOT being on the pump. They loved it.

Again, it was not until I met up with a former director of USACFA, Joan Finnegan Brooks, who told me how easy it was to manage her diabetes with a pump and that it made her life easier, that I decided to take the plunge and got one for myself. She had been using the pump for many years. What I appreciated about Joan's input was, like myself, she exercises a lot. I was concerned I would get too many lows with a pump. Using a pump, one gets small doses of insulin all day long (the basal rate) as opposed to what I was used to, only covering for carbs eaten. I was using Lantus at night for 24-hour coverage and short-acting insulin to control my glucose levels. But, the shots were getting tedious and painful. And with the pump, one can turn the amount of insulin down for a few hours while exercising or turn it up higher for times when one has an exacerbation.

So started my education on all things pump. I went to a class where I saw a twelve-year-old using a pump and loving it. I figured, at my ripe old age of 49, I could handle it. This old dog was ready for some new tricks.

It took a while to get used to putting the catheter on my body plus learning how to program the pump based on corrections from my educator. I went to two other training sessions. The educator input numbers and times of day for when insulin use

was higher and lower, the basal rate. She programmed my carb-to-insulin ratio for different times of the day and what my insulin-sensitivity was. Those things have changed a bit since I have now had my pump for about four months. Basically, I needed more insulin during certain times of the day. The good thing about the pump also is you can print out results of several weeks and see where the highs or lows occur and then fix amounts of insulin given based on what the educator suggests. It is process of learning and being open to input.

I just saw my endocrinologist and she was thrilled that: a) I was enjoying the new pump, and b) my control was good, even though I had been on a taper of prednisone and my numbers were a bit out of control for ten days, just recently.

Also, the meter that goes with my pump uses a radio frequency and it communicates to the pump. With the meter, I can send insulin to my pump without taking it out if it is lodged under layers of my clothing. I do usually clip the pump to my pants near where a belt would go. It looks like a beeper. Not that people use them anymore. But it is not that obvious. Most people think I am twiddling with my cell phone at meals and do not realize I am inputting numbers and taking insulin.

After inserting the catheter, I barely feel it. It is so much easier to give myself a correction bolus just by plugging in my blood glucose (BG). Something I rarely did when I took injections, sad to say. When I check my BG and it is too high, the meter or pump suggests how much I can give myself to lower to a better range. I then program in an amount and usually within two hours, I am within my comfort zone. The pump has many features and one is for when one's BG is high, as described above, but it also allows for inputting carb amounts plus current BG to tell you what it recommends for a bolus before a meal. I then decide what to put in.

This change to the pump has been positive and empowering for me. I feel like I have more control even though I am checking blood sugars more frequently. It feels good to say good-bye to syringes and hello to better health. But I would never have gotten to this point without the help of my CF mentors. Here's to all those who take it upon themselves to selflessly help and support us with our CF-related problems. Thank you for being there!▲

Andrea is 49 and has CF. She is a Director of USACFA and is the Executive Editor/WEBmaster. Her contact information is on page 2.

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Hearing Loss from Popular Cystic Fibrosis Antibiotic Could Be Treated with Vitamin Supplements

Gentamicin belongs to the amino glycoside class of antibiotics and is used to treat mostly gram negative bacte-rial infections. Side effects caused by antibiotic use are a major cause of concern among those with diseases such as cystic fibrosis. Nephrotoxicity and ototoxicity – a buildup of toxicity in the kidneys and ears, respectively – are the main side effects that doctors are most concerned about when prescribing these antibiotics. Gentamicin toxicity is the most common single known cause of bilateral vestibulopathy – a condition that occurs when the balance portions of both inner ears are damaged – which can lead to loss of hearing. It's worth noting that duration and dosage of these antibiotics is a major factor contributing to toxicity. A recent press release from the University of Florida reveals that around 2% to 25% of the people taking gentamicin experience hearing loss, but these surface only after a period of regular usage. Research has shown that use of dietary supplements loaded with antioxidants, beta-carotene, vitamins C and E, along with magnesium, have the potential to reverse the side effects in guinea pigs. Since the dietary supplements given in this experiment are not toxic and feature proven health benefits, they can be used effectively and safely on humans. This antioxidant-rich supplement quenches the free radicals produced by the antibiotics and hence protects against the side effects. What remains to be seen is whether this nutrient supplement reduces the efficacy of the antibiotic as a whole or not. If proven beneficial, these vitamin supplements could be prescribed as proper supportive therapy for infections needing gentamicin for treat-

Continued on page 14

Meet A New USACFA Director

Hello everyone, my name is Karen Vega and I am honored to have been elected as a new Director of this wonderful organization.

Having been diagnosed in 1981 at nine months of age, I am fortunate to have a mother who doesn't believe in statistics. She encouraged me to be involved in sports, especially soccer and swimming, and I had a relatively healthy childhood.

I am now 33 years old, living in New York and am currently a stay-at-home mom to my three young children. Prior to this, I earned my Bachelor's degree in Accounting from Fordham University and worked at a Wall Street accounting firm for eight years. In 2004 I married my husband, Alex. I gave birth to twin boys in 2008 and a daughter in 2012. My children definitely keep me busy all day long and are my motivation to succeed.

KAREN VEGA

In the past few years, I have become more involved in the CF community and have met some incredible people along the way. I serve as U.S. administrator and moderator of a Website and Facebook support group for women affected by cystic fibrosis who are already mothers or who are trying to become mothers through pregnancy, surrogacy or adoption.

In 2010, I became involved with the Boomer Esiason Foundation (BEF). I have become passionate about exercise and am a big advocate for it in the CF community. I ran in the NYC Marathon for Team Boomer in 2010, and this past March I ran the Rock CF Rivers Half Marathon. I continue to make exercise one of the priorities in my life and I attribute my good health to it.

I look forward to contributing to *CF Roundtable* and working with USACFA. ▲

TILLMAN *continued from page 13*

ment. More research needs to be done on this before one can move to clinical trials and get the same approved officially.
http://tinyurl.com/nf3hnxy

Bacteria in Cystic Fibrosis Lung Infections Become Selfish

Bacteria that infect the lungs of cystic fibrosis sufferers lose their ability to work together, becoming more selfish and less cooperative the longer the infection, say scientists. Researchers hope that better understanding how the potentially-fatal infection changes over time will make it easier to treat. During chronic infections, bacteria will change their behavior toward each other. The researchers looked at four different "cooperative" traits to see how the bacteria evolved during infections. These included production of signal molecules bacteria use to communicate with other bacteria called quorum sensing molecules; the concentration of protein-digesting molecules called proteases; the production of bright green pyoverdine, which is used to bind iron; and how good the bacteria were at forming biofilms. Biofilms are formed by groups of microorganisms in which cells stick to each other on a surface. They found that the longer the bacteria had been infecting the lung, the fewer signals they sent out to other cells, the less biofilm they formed and the less pyoverdin they produced. Understanding how these bacteria evolve, and why they choose certain behaviors over others, will make it easier for treatments to eventually be found.
http://tinyurl.com/kogx589

Arch Biopartners Enters Option to License University of Cincinnati Patent for Treating Bacterial Respiratory Infections

Arch Biopartners, Inc., announced that it has entered into a one-year option agreement with the University of Cincinnati to exclusively license the commercial rights to a U.S. Patent for treating bacterial respiratory infections using acidified nitrite. The mucoid form of Pseudomonas aeruginosa (Pa.) is a very challenging infection to treat due to its high resistance to both antibiotics and phagocyte-mediated killing. The technology developed by Dr. Hassett constitutes an innovative, non-antibiotic method for dealing with mucoid Pa. infections where the emergence of resistance is highly unlikely.

Continued on page 19

PHOTO BY STEPHEN BOYER

Hand in Hand

Surrounded by sickness
Together we stand
Kaeti and Allison
Hand in hand
One without the other
We're not as strong
Life doesn't make sense
Everything seems wrong
But with each other
At our sides
We are in control
And take great strides
To living life however
We desire
A thought to CF
But it's a mere flat tire
Because in the long journey
It's a small anecdote
But our lives are our stories
CF is the sidenote

-A. Best, 2004

"Through the Looking Glass: Images of Adults with Cystic Fibrosis" and "Caregiver Stories" are projects of Breathing Room, a non-profit organization. Breathing Room hosts these and other projects to facilitate open and candid communication in the CF community, supports the development of a community of adults with CF and provides education and insight for families, caregivers, and medical professionals who impact our lives.
To learn more about us and view more images in the collection, please visit our Website at:
http://www.thebreathingroom.org

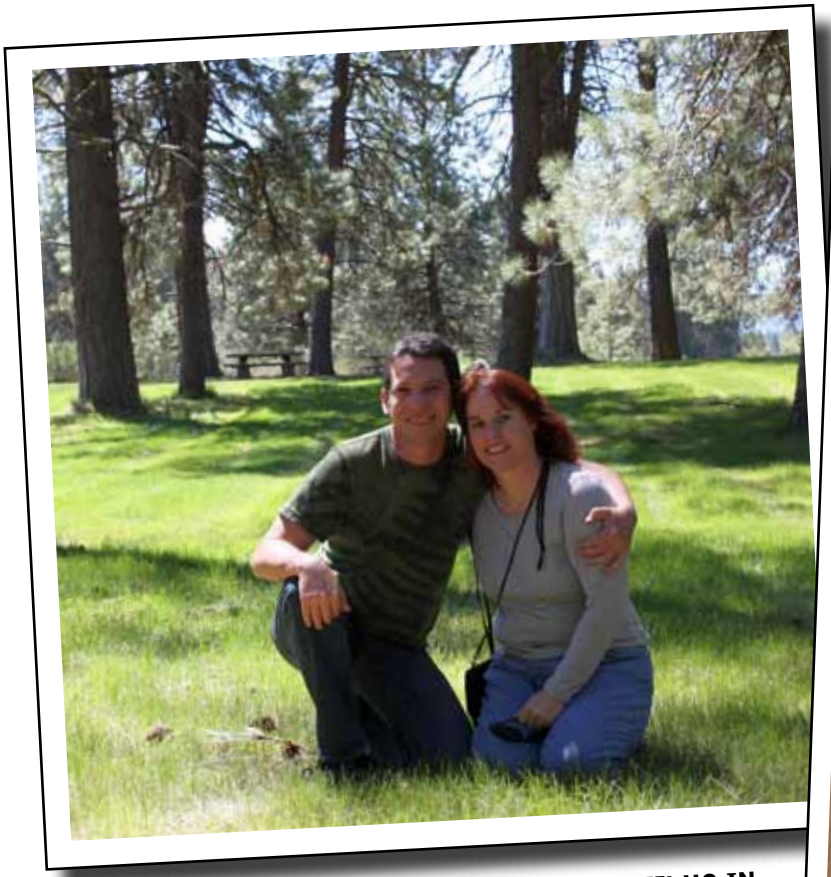
FROM OUR FAMILY PHOTO ALBUM...

MERANDA HONAKER IN EMERALD ISLE, NC.

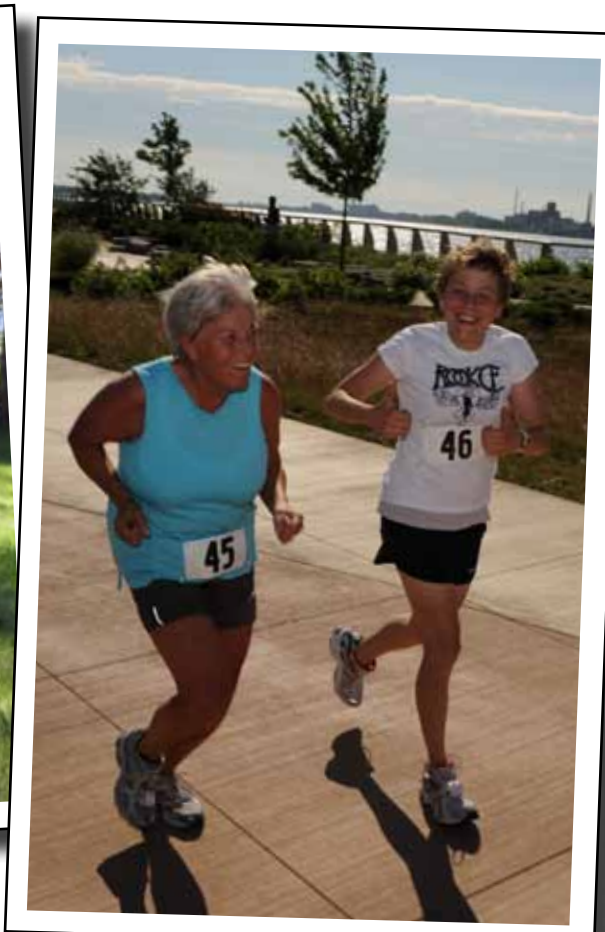
ANDREW BYRNES AND ISA STENZEL BYRNES IN WASHINGTON, DC.

DR. JERRY NICK HOLDING HIS WILLIAM J. MARTIN II DISTINGUISHED ACHIEVEMENT AWARD NEXT TO BETH SUFIAN.

KAREN VEGA AND HER CHILDREN, LEFT TO RIGHT, JAYDEN, KAELI, LOGAN.



TOM MARTIN AND GIRLFRIEND THERESA BOTEILHO IN OREGON.



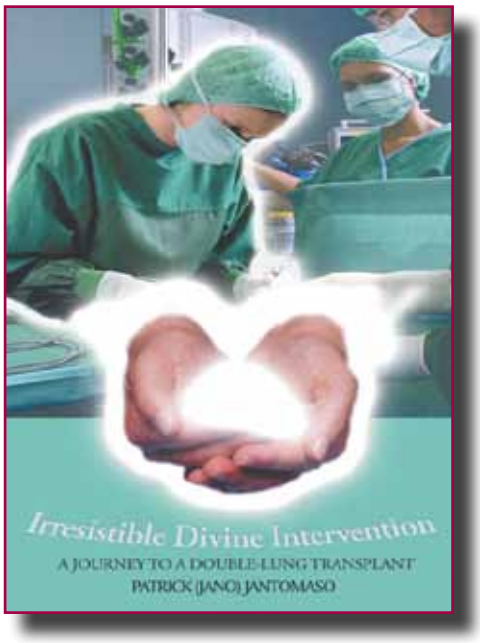
EMILY SCHALLER RUNNING A 5K WITH HER MOM, DEBBIE SCHALLER.

BOOK REVIEW

Irresistible Divine Intervention

Jon Jantomaso's journey to a double-lung transplant

By Patrick (Jano) Jantomaso
Published by Authorhouse, 11/26/2012
ISBN: 978-1-4772-7591-7
74 pages
Reviewed by Paul Feld



I believe I have done three book reviews for *CF Roundtable* in the past and have read about a half dozen books about people with CF who have had bilateral lung transplants. I also know, personally, at least 25 good friends who have had the life-extending procedure done. I have CF and had a double-lung transplant, as well, 10 years ago. Since the author's son Jon and I are only nine years apart in age, and we both had our transplants in our 40s, I was excited to read about his journey. This is, by far, the most challenging book review I have ever written.

When I was asked to do this book review, I knew that the author, Pat Jano, was writing about his son, Jon, and his experience with CF and transplant, and I had anticipated another quality CF success story. It turns out my expectations were dead wrong. This 74-page book is really a four-hour (the time it took me to read it) Christian sermon, and Jon's story, or lack thereof, gets lost in the preaching. In an accompanying letter to me that came with the book, Pat Jano said his purpose for writing the book was simply to raise money for CF. I sit here still wondering how hundreds of bible verses of the Christian faith can serve that purpose, as opposed to expressing the extreme challenges faced by people with CF every single day, before and after transplant. It seems to me, when people get a grasp of the challenges, they would more likely donate for the cause.

Now, back to the book. Knowing the author's background a bit further, you may better understand the direction the book takes. Patrick Jantomaso is an ordained pastor/minister and this is his fifth inspirational book. He is extremely well-versed in the bible and has a unique ability to pull out very specific chapters and verses to get his point across as it relates to everyday happenings. If you are of a Christian faith, as I am, much of what he writes will resonate with you on a spiritual level and, hopefully, strengthen your relationship with God. If you are not of a Christian faith, your interest in this book will wane quickly as you search for information on his son, Jon, and CF. There may be four to five pages about Jon and CF in the entire book. It left me clamoring for so much more.

I would recommend this book to devout Christians who emphatically believe in blessings and miracles and are continuously optimistic about their relationship with the Father, Son, and Holy Spirit. For others, I think that you would have to enjoy a four-hour sermon to enjoy the book, and I believe few people fall into that category. ▲

Paul Feld is 57 and has CF. He is a Director of USACFA. His 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 Facebook support group, please e-mail Karen Vega at: kvega@usacfa.org.

MARTIN *continued from page 1*

tioning, not including the exercise, but even more so if you have CF lungs – depending upon the severity of your disease. I need the extra calories and energy because of my vigorous exercise routines; I mountain bike three times a week between 1 to 2 ½ hours, and lift weights at the gym two to three times a week. In Aptos there is a great staircase by the beach (Seacliff Beach). It's a great place to work out with cool salt air to breathe, and I count 151 steps from top to bottom. I usually attempt three or four reps of the "Aptos Stairs" at least once or twice a week.

I see a plant-based diet as leverage to fight against chronic illness and susceptibility to catching viruses such as H1N1 among many other ailments. According to the research of Dr. John McDougall, the evidence overwhelmingly shows that a vegan diet can save you from all these everyday ailments that we have to deal with. Dr. McDougall has been researching and studying the effects of not eating a vegan diet for decades and discovered the benefits of eating a plant-based diet in the process. He and many other medical doctors, including T. Colin Campbell, a biochemist known for authoring the book, *The China Study*, have examined the relationship between the consumption of animal products (including dairy) and chronic illnesses such as coronary heart disease, diabetes and cancers of the breast, prostate and bowel. The author concludes that people who eat a whole-food, plant-based/vegan diet – avoiding all animal products, including beef, pork, poultry, fish, eggs, cheese, and milk, and reducing their intake of processed foods and refined carbohydrates – will escape, reduce or reverse the development of numerous diseases. Since I'm a transplant patient and immunocompromised, I'm at high risk for cancer among many other ailments. Even though I did put off my colonoscopy far too long, being middle-aged with CF, I'm happy to report my recent colonoscopy is negative (no evidence of cancer or polyps etc.).

> "My health success has come from listening to the experts and contemplating for me what I believe is best overall."

With all that said, I think having a healthy skepticism about what others write and study is wise. I take the evidence and information from others, then apply it to my situation. Since these books from McDougall and Campbell apply to all people, I have to consider my CF condition as a huge factor. So I do eat more oil and fat (plant-based) than what is recommended by these books. My health success has come from listening to the experts and contemplating for me what I believe is best overall. I've been vegan for one year now and so far the results are very good and I'm feeling great! I'm eight years post-transplant and 46 years old, all my numbers are very good and I'm happy to report no more GI-related pain issues.

I consider myself lucky to belong to a regular weekly riding group that we call "Tuesday Night Fever." We all mountain bike all year round, down to 40 degrees in the winter and up to 90 degrees in the summer, although rain is a no go for us. We have powerful bike lights for winter darkness. That's my story (plant-based diet) and I'm sticking with it.

Thanks, and to your health! ▲

Tom is 46 years old and has CF. He is eight years post-bilateral lung transplant. He can be followed on STRAVA, a GPS app for cyclist and mountain bikers. His e-mail is: tbmartin12@yahoo.com. He can also be followed on Instagram at http://instagram.com/tommartin600, where he posts photos of nature trails and scenery while mountain biking and of vegan food that he loves to eat. His mountain biking video can be found at: https://www.youtube.com/user/tubemartin600/videos.

TILLMAN *continued from page 14*

http://tinyurl.com/l9vlyub

Cystic Fibrosis, COPD Bacterial Respiratory Infection Treatment Being Developed by Arch Biopartners, University of Cincinnati

Arch Biopartners has entered into a one-year option agreement to license the commercial rights of a University of Cincinnati-developed technology for treating bacterial respiratory infections associated with diseases such as cystic fibrosis. The novel treatment utilizes acidified nitrite, a non-antibiotic method. During the one-year option, the company will be working to assess the potential and logistics of conducting a Phase II clinical trial. The goal is to test how effective the UC technology can be when bacteria

Continued on page 21

WELLNESS

Well This Is Awkward

By Julie Desch, MD

This issue of *CF Roundtable* focuses on conditions that are "part of CF," and I suspect there will be much written about CFRD, bowel obstructions, and perhaps even some "waiting for my transplant" stories. But I want to talk about the very large elephant in the room. Or I should probably say "elephants," as there are a few.

Maybe it's our culture or maybe it's just me, but it seems to me that CF is a disease that comes with more than its fair share of embarrassing accompaniments. Obviously, there is the cough that turns heads (especially in the gym), which leads to the oft-asked questions regarding possible contagion or history of smoking. I think we all get used to this and eventually come to expect it without a second thought. Some of us (or again, maybe its just me) like to play with the tendency of people to freak out upon hearing the nasty sounds coming from our mucus-laden lungs. Occasionally, I'll answer the contagion question with, "Don't worry, my doctor told me I could leave the house as soon as the rash disappeared," and then walk away. This shuts them up for awhile, and it really clears out the wait lines for the weight machines.

But, there are some issues that nobody can truly get used to. The main one for me has to do with the CF gastrointestinal tract. I refer to this as the odeur de CF. Let's face it, it is often the case that when we leave a bathroom, a hazmat team should probably be summoned. I can't count the number of times that I, as a young child, would linger hiding in a public toilet stall, waiting until every last person was gone before I would venture out, head down, practically running out the door. If someone happened to enter before my great escape, I would nod empathetically at the look of horror on her face, and gesture to the stall I had just left, telling her that whatever she did, she should NOT go in there, as I was sure something had just died. You improvise as you need to.

In medical journals, the cause for such disgust is referred to as "malodorous stools." I always chuckle at this, as it's a bit like calling NFL football a "rough" sport. When a dog—you know, the creature that gets acquainted with another by sniffing its butt—won't even enter the bathroom, you know it's bad.

Aha, I have heard of a product called "Just A Drop." Supposedly, when one puts a drop of the magic potion into the toilet bowl prior to… evacuation…there is absolutely no smell and you can prance out of the restroom without a care in the world. That's right, that is what they say. I have since ordered said product and just today tried it for the first time. I believe that the active ingredient is some kind of mint oil, perhaps eucalyptus, mixed with other mysterious substances that smell vaguely perfumey and disinfectant-like. According to the packaging, the magic oil forms a barrier at the top of the water, through which no malodorous fumes can escape.

> *"Let's face it, it is often the case that when we leave a bathroom, a hazmat team should probably be summoned."*

JULIE DESCH, MD

Drumroll please: It sort of works! There is a small caveat, however. The oil is a tad effervescent when mixed with the water in the toilet. If you have…sensitive mucosal membranes…you may feel a slight burning sensation in a place where this would seem unusual. It takes some getting used to.

A closely related problem is intestinal gas, or flatulence, if you will. The truth is that everybody farts, even those with unblemished CFTR. In fact, it is said that the normal number of farts per day (FPD) is 15. I don't know how that number was arrived at, but I choose to believe it and not question the Google. I know people who swear

that they *never* have the toots, and they are liars. Apparently, people can fart without even knowing it…little microfarts if you will. So when you hear someone say, "I don't fart," tell them that you know for a fact that they do, at least 15 times a day.

But I digress. Microfarting is not our issue now is it? Zillions of bacteria living in our colons feast on the partially undigested food that is constantly provided to them, thanks to insufficient pancreatic enzymes. The byproducts of this bacterial gluttony are CO_2, methane, hydrogen gas, and even the dreaded sulphur. This is not just offensive to nearby noses, it can be downright painful, as I'm sure you know. Again, I am brought back to my childhood, when I would stubbornly not permit my body to do what it desperately needed to do on school property. As a result, I would be in nearly constant pain during school hours, dreaming of being able to stick a big needle in my belly to relieve that pain, just as I had read that Dr. James Herriot had done to the cows with "the bloat" in *All Creatures Great And Small*.

Consulting the Google again, I learned that there is a possible course of action we can take, at least when it comes to the smell. Our freedom comes in the form of "charcoal pads" one can insert in one's underwear. Allegedly, the charcoal will absorb the smell of any wayward bacterial byproducts, and you will go merrily on your way with nobody the wiser. You're on your own when it comes to dampening the sound. Good luck.

I would be remiss if I didn't mention another issue that, sooner or later, we will all have to wrestle with. Given the laws of physics in our universe, there will come a day when tissue that normally resides inside the body will come to rest in an abnormal and uncomfortable position. I'm talking hemorrhoids here. Given the hours upon hours straining on the toilet (do you sense a theme in this article?), plus the tens of millions of valsalva maneuvers in the form of coughs, the very vascular lining of the colonic caboose (the anus) wants to get the heck out of Dodge and never come back. Sometimes, if you have really bad luck, the whole dang rectum says "wait for me," and you get rectal prolapse. None of this is fun or funny, and often surgery is required.

Finally, and I'm predominantly talking to you women out there in CF-land regarding this problem, can we speak a minute about urinary incontinence? It's called "stress-incontinence" because it's not like we are just randomly peeing our pants. It happens when we cough…which we do…a lot. Take heart, we are not alone. Women who have gone through many pregnancies often struggle with this issue as well. And if you have CF and have given birth, God help you.

Doctors tell you to do Kegel exercises, which are like bicep curls for the muscles of the pelvic floor. "Pretend like you are stopping a pee in midstream," they say. Do this over and over, multiple sets of innumerable reps, throughout the course of the day. Do it standing in line at the grocery store. Do it at red lights. Do it during commercials when watching television. Do it every time you have to change your underwear because you peed your pants laughing again.

It doesn't work. Fortunately, we can repurpose sanitary napkins until the day comes when we really do need Depends. Just nestle it right in there with the charcoal pad and you are good to go. ▲

Julie is 53 and is a physician who has CF. You may contact her at: jdesch@usacfa.org.

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are resistant to antibiotics. The Phase I results revealed that the new drug is safe at or below the maximal tolerated dose.
http://tinyurl.com/kdyv269

New Cystic Fibrosis Clinical Trial to Be Launched by Kamada Ltd. This Summer

Israel-based Kamada Ltd. indicated recently in a press release that their lead drug candidate Alpha-1 Antitrypsin (AAT) will be tested in a new round of clinical trials for cystic fibrosis this summer. Kamada indicates that the company plans to initiate a U.S. phase 2 clinical trial of its inhaled AAT to treat cystic fibrosis in the second half of 2014. The company has had previous success in researching and developing cystic fibrosis treatments, including Bramitob, which features Tobramycin as its active ingredient and offers "management of chronic pulmonary infection due to Pseudomonas aeruginosa in patients of 6 years and older with cystic fibrosis."
http://tinyurl.com/mxspjtn

Xenetic Biosciences Announces Positive Phase 1 Clinical Data for PulmoXen(TM) for Treatment of Cystic Fibrosis

Xenetic Biosciences, Inc., announced the positive results from its Phase I clinical trial of PulmoXenTM for the treatment of cystic fibrosis. In this First-in-Human study, PulmoXen

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COUGHING WITH A SMILE...

Keep On Keeping On

By Jennifer Hale

Hello *CF Roundtable* readers! Hope this issue finds you all doing well and getting ready to enjoy the summer. I know those of you up North could really use some warm sunshine on your face! The topic for this issue is: "Dealing With Conditions That Are Part Of CF." The running joke in my house is that if something is hurting, not going right or whatever, it's CF related! But the not-so-funny thing is that it probably *is* truly CF-related. What everyone is learning now with CF people living longer is that the disease is a full-system attack on all parts of our bodies and all functions of our bodies.

When I was first diagnosed at the age of two, CF was all about the lungs and pancreas. Now we have problems like CF-related diabetes, arthritis, GERD and many other complications. I often wonder how many different conditions I can handle all at once! I am sure if you have been following my column, you know I am the type of person who says, "Bring it, I can take it!" Although I am a positive person, it is not easy living with CF or, as I tell my husband, it is not easy "being green!" Thank you Kermit the Frog for that one!

The two problems that I am currently dealing with are my CF-related diabetes, also known as CFRD, and low lung function. Low lung function is not really a condition or another disease from CF but just a problem that occurs due to the nature of the disease as it progresses throughout my body.

CFRD has been challenging to say the least. I was diagnosed in 2005 and I have to say it is such a high maintenance problem to have on top of the CF. I am constantly checking blood sugars, loading up on carbs before doing something physical, carrying around juice packs etc. The difficulty with CFRD is when you are an active person, as I am, blood sugar is constantly being challenged. To make things easier for me, I always have juice packs on me, in case I have a low blood sugar event. I think I need to buy stock in Capri Sun soon! But when that sugar drops, you can not fool around and you have to treat the low immediately. So you will always find me with juice packs in every purse, bag or cooler. I guess the good part to having CFRD is I can always bring drinks and granola bars into stadiums or airplanes due to having diabetes. But all in all, I do the best I can to keep my HgbA1c at a good level. I test my sugars constantly and try to do the best that I can. That is really all you can expect from yourself, do the best you can given the situation you are facing.

> *"What everyone is learning now with CF people living longer is that the disease is a full-system attack on all parts of our bodies and all functions of our bodies."*

JENNIFER HALE

Low lung function is plaguing me and I am not a happy camper! In four years I have dropped from an FEV_1 of 65% to a steady 38-41%. Let me tell you it has not been easy going through this downturn and trying to live with a low lung function. I now use oxygen when I work out at the gym and, yes, I still go there to work out even though it is so very hard. I am out of breath doing even just menial things. But, as I like to say, it is what it is.

I notice for the first time in my life that I look at others running and I am envious. I am envious of those who can walk around the block and not be overly out of breath. I am envious of people at the gym who are working out really hard and doing these great moves for cardiovascular, but I cannot do those. Think kettlebell swings! I have been active my whole life. I have played tennis, baseball, run on the treadmill, taken spin classes and swum. And now it is a different story for me due to the inability to breathe.

I have also been a swimmer my

whole life. I have never been scared of the water because I always knew what a strong swimmer I was. But for the first time recently I actually got scared I was going to drown. I started to swim out to a sand bar. About six breast strokes in, I could not breathe at all even though I was taking huge rapid and deep breaths. So I turned around and was just focused on getting to where I could stand. Well, I made it and I have never been that out of breath in my life! I guess a lung function of 38% does not allow one to swim. So I learned the hard way. It was very scary and very sad because I no longer can swim. The girl who grew up swimming all summer long, the girl who could jump from a boat and swim around, the girl who snorkeled in Aruba and Hawaii. No longer am I a swimmer. Do they make adult water wings? LOL!

It is easy to give up being active when it is so difficult to be active. I actually feel better during the day when I do not work out. But with that said, I will NEVER give up my work- outs and my activities. I have just made A LOT of adjustments. I wear my 0_2 when I go the gym and I even leave it on a little longer when I am done, to give my lungs time to recoup. It seems to be helping me to not feel so tired or feel sick at the end of the day, when I leave my 0_2 on longer after working out. For now that is my new method to get through my workouts.

As far as other activities, I have had to just modify. I have my husband park the car closer to where I'm going, have him go get the car when we are out, or use my 0_2 if I know I will be active outside the formality of the gym. I know working out is so good for your lungs, and I feel small steps are better than no steps at all. I believe just moving, no matter how intense or not, is the key.

I believe in consistency; consistency in working out and not so much how hard the work out is but that I am consistent in doing something four or five times a week. Really, all I can do at this point is to adjust those sails and go with the flow. This is how I handle the low lung function. I keep moving and doing the best I can for each day. It is not about how hard you work out but how consistent you are in getting out and moving and doing something. That is just my opinion.

I have also come to realize that I cannot compare myself to others who have CF. Everybody has different lung function, severity of symptoms and other conditions that arise due to CF. So, don't compare yourself to others, just set goals that YOU can achieve. Use other people's ways of doing things to motivate you to do the best that you can in YOUR life and YOUR situation.

I leave you with this quote from Tom Cruise in Vanilla Sky, "Every passing minute is another chance to turn it all around." Get out there and keep turning it all around with each passing day, with each passing minute! ▲

Jennifer is 42 and has CF. She and her husband, Mark, live in St. Petersburg, FL. You may reach her at: jhale@usacfa.org.

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was administered to 12 healthy volunteers via inhalation daily for seven days, in two doses of 2500IU and 5000IU, and was found to be safe and well tolerated. PulmoXen is a novel, modified form of recombinant human DNase I (rhDNase I) designed to be a next-generation version of Pulmozyme®. PulmoXen is a polysialic acid – conjugated form of rhDNase I that is being developed using Xenetic's patented PolyXen® delivery technology, with the intent to reduce the required dosage frequency by enhancing the stability and pharmacodynamic profile of the enzyme in sputum. A comparative evaluation of PulmoXen versus Pulmozyme, using ex vivo models of cystic fibrosis sputum, confirmed superiority of PulmoXen with respect to stability and enzymatic activity when digesting DNA in sputum from cystic fibrosis patients.
http://tinyurl.com/ny6yc34

Gilead's AZLI Repurposed to Treat Burkholderia in Cystic Fibrosis Patients

Another cystic fibrosis Phase 3 clinical trial from Gilead Sciences has been completed, with goals of determining the safety and efficacy of aztreonam for inhalation solution (AZLI) in cystic fibrosis patients with chronic *Burkolderia* species infection. AZLI is an inhaled antibiotic which is administered via nebulizer. The mechanism of action of aztreonam involves binding to certain bacterial proteins to disrupt cell wall synthesis, leading to bacterial cell death.
http://tinyurl.com/qadr8xu

Vertex Says Cystic Fibrosis Drugs Shown to Boost Lung Function

Vertex Pharmaceuticals, Inc., said a combination of its cystic fibrosis drug Kalydeco and an experimental compound was shown to improve lung function in a mid-stage trial. The study

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IN THE SPOTLIGHT
With Emily Schaller

By Andrea Eisenman and Jeanie Hanley

We were asked to interview Emily by a director of USACFA, Beth Sufian, after learning of Emily's amazing work. She founded a nonprofit for CF and speaks around the country about living with CF. Starting out as a drummer for an all-girl rock 'n' roll band, Emily decided to organize a CF fundraiser in Detroit featuring local bands and went on to establish the Rock CF Foundation. Drumming cleared her lungs so well that she caught the exercise bug and decided to try running, starting very slowly and progressing to half marathons and Iron Man competitions. Emily is so energetic, even talking to her over the phone made us want to get out there and run. She travels around the nation as a motivational speaker and promotes CF awareness primarily through organizing running events. Check out her Website at: www.letsrockcf.org for the Rock CF Foundation and consider participating in one of her events and enjoy her great laugh, too. Please welcome our next star. Spotlight please!

EMILY SCHALLER AFTER COMPLETING THE ROCK CF RIDE, FROM WASHINGTON, D.C., TO PITTSBURGH.

Age: 31

Diagnosed with CF: at 18 months of age.

Occupation(s):

President/CEO of the Rock CF Foundation and Freelance Race Director

Where did you grow up?

An island in the Detroit River called Grosse Ile, MI.

Any siblings?

Two older brothers.

What was your family life like?

My family is hilarious! For us, laughter was truly and still remains the best medicine. We never made CF the center of the world; we had no time for that. Instead we lived life and had fun!

What were your symptoms as a child?

Chronic ear infections, colds and such, but most of my symptoms were in the digestion department, i.e., no weight gain. I was diagnosed with failure to thrive before finally being diagnosed at 18 months.

What would you say is or are your greatest accomplishment(s) so far?

Personally, I think finishing two thirds of a half-iron distance triathlon. Confusing I know, but I cycled 56 miles and then ran 13.1. Although I grew up on an island, I literally don't know how to swim. Luckily my friend (who also has CF) knocked the 1.2-mile swim out for me that day.

Professionally, I am super proud of the Rock CF Rivers Half Marathon race that I (and my committee) established in 2011. With 2,000 runners from over 15 states and Canada, this race is not only the Rock CF Foundation's signature event, it is also a top-notch and nationally top-rated race.

What is the Rock CF Foundation?

The Rock CF Foundation is a 501(c)(3) nonprofit corporation that I officially founded in 2007. Rock CF started with using music (live and recorded) to raise funds and awareness for CF. Rock concerts and CDs were what kicked us off. As my personal focus shifted to running, cycling and an active lifestyle, we established this mission: to increase the quality of life for people with cystic fibrosis. With the help of a core group of volunteers, the Foundation utilizes the arts, entertainment, fashion and fitness to support research initiatives and heighten public awareness in the fight against cystic fibrosis.

Why did you start it?

A few things led to me create Rock CF. I felt that the 18- to 35-year-old public was being left out of the CF equation when it came to awareness. There were the CF walks and other fundraisers that attracted CF families and friends. Then there were the upscale black tie events and golf outings that were geared toward higher income families and corporations. There weren't many fundraising and awareness outlets that reached the everyday, healthy 18- to 35-year-olds; or as I see it, the next generation of parents who could be affected by CF. I wanted to change that. The concerts brought local Detroit bands and their fans together for one night of rock and roll. This was a captive audience that gave us the chance to tell people about CF while raising some money for research. It was great!

Also I wanted "Rock CF" to be a positive movement. CF can be ugly but sometimes we just need a little bit of confidence to fight on. Rock CF is empowering! The Rock CF merchandise gives people (with and without CF) a chance to wear their heart on their sleeves when they wear one of our cool shirts, hoodies or hats!

How many volunteers work for you?

Anywhere from 1 to 3 to 300 for the half marathon race!

Are they an unruly rock 'n' roll group? We have some unruly rockers for sure. That's what makes us tick.

Is Rock CF nationwide or international?

Over the past year we have really become internationally known. It still blows my mind! E-mailing and messaging daily, and shipping merchandise to Australia, New Zealand, Jerusalem, Europe and all over the USA every day.

What's the biggest event you've had so far?

The Rock CF Rivers Half Marathon on Grosse Ile is not only our signature fundraising event but it is also our biggest single day awareness movement. Runners from 15-20 states fly in from all over the country and Canada for the race. We should see over 2,500 this year! Also all CF patients can run or walk any of the three distances for free! Anything I can do to encourage exercise!

Where do the proceeds go?

Rock CF supports the mission of the CF Foundation, and we donate proceeds to help fund its research initiatives. My big thing is awareness and making CF mainstream. The Rock CF online shop allows us to sell super cool and stylish clothing that everyone wants to wear, whether they know what CF is or not. The sales of this merchandise then is fueled back into the Rock CF Foundation and the CFF. We also have in the works the launching of a new program that has to do with running shoes and CF patients… stay tuned for that!

Tell us about the "Virtual CF" events.

The virtual runs are coproduced with my friends at CysticLife.org. Currently we organize two runs a year (one is on Thanksgiving) and this is our chance to bring the whole world together on one day to do something good for ourselves while supporting CF with people all over the world.

How often do you participate as a speaker for CF events and where do you go?

Usually the spring and the fall are the busiest for me. CF Care Centers host their Family Education Days and I'll drop in as a speaker for that. I also speak at CFF events around the country, too, more gala-type events that are a blast! I've spoken in around twenty states and really love it!

How did you learn to play drums and what effects did playing with a band have on your health?

I got my first drum set when I was about five and then another one in third grade. These were really just to bang around on and I never had lessons or really learned how to play. When my friends and I decided to start a band in 2004, that's when I got another drum kit and started learning how to play. I would just strap on an iPod and start hitting away. Then we started writing our own songs and the rest is history. When I first started, I could really play for only a few songs at a time because it made me cough so much. It was literally the most exercise that I was getting for those five years after. It got easier the more I played, but I think playing drums kick-started my path to exercise.

How was it to reunite with your band for the December 2013 event?

The show was great! After not playing in three years it was great to get on stage with my friends, play some rock and roll and raise some money and awareness for Rock CF!

What led you to start running for exercise?

Simply put, I was sick of being sick. I knew that there was more that I could do for my health than just doing my meds, vest and treatments.

How long could you run in the beginning and how long did it take to be able to participate in half marathons and Iron Man competitions?

When I first started I could barely run one block. I had to run one block or to a landmark, walk for a little bit and then run. It was really hard when I first started. It took about three to four months to build up to running a 5K. From there it was another six to nine months and I was running my first half marathon.

Was running better than playing drums?

No doubt running did more for me than playing the drums. But when I was doing no other exercise at all, drums were an okay place to start.

What are your CF genetic muta-

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tions and when did you start Kalydeco?

G551D & 1078 delT. The secret is out! I've been on Kalydeco for about three years now. I was on phase 2 and 3 of the clinical trial and also the open label portion.

Since starting Kalydeco, what differences in your health have you seen?

It's hard to put into words what I have experienced. On paper it looks good, "lung function increased, weight gain, drastically lowered sweat chloride levels, etc." But I have to say that I have never felt better in my entire life. It's like someone flipped a switch. Most days my cough is absent. I can take a full deep breath now without coughing. I can laugh without coughing. When I run or cycle my sweat is way less salty. This all sounds good too, but until you experience what I am experiencing, nothing I'm saying does this justice. I still do my daily routine of treatments, vest, enzymes, pills and inhaled antibiotics. And I have needed IVs since starting Kalydeco, but my day-to-day life is unbelievable.

Does using Kalydeco replace your running exercise?

Nothing will ever replace running. I'm a runner! Kalydeco just makes running so much easier, which means I have to push myself to go faster and/or farther now!

What did it feel like to turn 30?

Turning 30 was kind of scary. I don't know why but it was just an odd feeling. Now that I'm 31, I don't even care.

What has been your most difficult challenge?

The hardest thing for me as I get older is losing my friends with CF. It never gets easier, especially when I can benefit from Kalydeco and most of my friends can't. This is hard.

What gets you through the tough times?

Lots of yoga, running and exercise. This grounds me and helps me to realize that whatever I'm going through may be tough, but I can get through it. Also humor gets me and my family through anything.

For you, what are the benefits of having CF?

I wouldn't be who I am today if I didn't have CF. Living with a chronic illness builds so much strength and character in people. It gives you a drive that you don't see very often in the average person. I wouldn't be where I am right now if I didn't have CF. I have been fortunate enough to travel the country, see amazing things, meet amazing people and CF patients/families. I love my job and everything I do. If I didn't have CF, I might be working some 9-5 job that I absolutely hate that does nothing to benefit others in a positive way.

What is your funniest CF moment?

I would say that all of my hospital stays at Children's Hospital in Detroit were super comical. Lots of memories from that place. I'm not going to give anything away, but one moment involves me getting a cast on my foot at 3 am…and my foot was not broken.

What or who is your favorite music, singer or band?

Brandi Carlile, hands down.

Knowing that your family founded a Lutheran Church in Michigan, is that why you're obsessed with Germany? Where is(are) the best place(s) you've travelled to internationally? Any CF issues while travelling?

My family didn't found *the* Lutheran church, but they did establish a Lutheran church in Flat Rock, MI. My ancestors on both of my parents' sides were from Germany, so I grew up attached to Germany. I love it! I took German in high school and try to speak it whenever I can, especially if I meet someone who speaks German.

My best friend from childhood lived in Amsterdam for a few years, so I was lucky enough to visit her a couple times. We travelled to Berlin, Cologne/Koln, Paris, Brussels, Rome and Barcelona. Amsterdam is one of the greatest cities in the world. So bike friendly and it just has a great vibe. Luckily, I had no international travel issues. Because my friend has known me since we were six, she was able to perform manual CPT while I was there and I didn't need to bring the vest.

What goals are you currently working toward?

My goal is to run a half marathon in all 50 states. So far I have five done, I'm not good at math but sounds like I have 45 to go. I'll be old and gray by the time I reach that goal…but at least I'll be old and gray!

Anything you wish you could do over again?

I wish I could relive my early 20s again. I would have kept exercising out of high school and stopped eating fast food!

Do you have a message for those with CF who don't exercise or who feel they can't?

Running, biking, yoga, rebounding, Zumba and exercise in general is not easy when you are just starting, especially if you have CF. Find something you like to do, and do it! My advice is to start slow, like really slow. Don't give up when it's hard, just slow down. Keep track of your exercise each day so if you do get discouraged you can look back and say, "I can… because I did! Yesterday I walked one mile!" Trust me, some days are way harder than others, but you just have to want to do it. Some days it's not going to be as good or as far as you planned, but you still will do it! ▲

Jeanie Hanley is 51 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2. Andrea Eisenman is 49 and has CF. She is a Director of USACFA and is the Executive Editor of CF Roundtable *and Webmaster. Her contact information is on page 2.*

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

USACFA Congratulates Jerry Nick, MD

Jerry Nick, MD, was awarded the 2014 William J. Martin II Distinguished Achievement Award on May 21, 2014, at the American Thoracic Society (ATS) International Conference in San Diego. Dr. Nick is the Adult CF Center Director at National Jewish in Denver, Colorado.

The award is given to a clinician who has shown outstanding commitment to the well-being of people with chronic respiratory disease and contributed to medical research in pulmonology. There were ten other nominees who were specialists in the treatment of ten other specific respiratory diseases. Dr. Nick was chosen for the award in recognition of his almost 30 years treating adults with cystic fibrosis and conducting ground-breaking medical research in CF.

JERRY NICK, MD

Dr. Nick is Director of the largest adult CF center in the world with more than 300 patients. He heads a staff of more than 15 and sets the tone for a CF Care Center that makes sure a patient who needs care is seen even it means the CF Center is seeing patients when it is dark outside.

The ATS was impressed with Dr. Nick's ability to develop proof of concept research that is innovative and cutting edge.

USACFA salutes Dr. Nick and congratulates him on being presented with this prestigious award. ▲

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found that treatment with Kalydeco and the experimental drug VX-661 for 28 days resulted in a 4.6 percentage point improvement in mean lung function for patients with two specific genetic mutations. The most common side effects seen during the Phase 2 trial included cough, headache and upper respiratory tract infection.
http://tinyurl.com/p3nxy5p

OPKO Acquires Inspiromatic Dry Powder Inhaler, Potential for Cystic Fibrosis Treatment

Inspiro's Inspiromatic™ is a dry powder inhaler that can deliver small or large molecule drugs to treat respiratory diseases. Dry-powder inhalers are an attractive alternative to nebulizers to deliver cystic fibrosis treatment. It is more convenient and time-efficient for patients to inhale dry-powder medication than wet aerosols. Nebulizers need a compressor unit or pressurized air if they are jet-type and electricity if they are ultrasonic. What's more, only approximately 1-32% of the intended dose reaches patients' lungs when delivered via nebulizer. Studies have demonstrated that at least 40% of the intended dose from dry-powder inhalers is emitted, and the duration of administration of medication can decrease from at least 20 minutes to less than a minute.
http://tinyurl.com/mo8hurz

Positive Data on Pulmatrix's iSPERSE™ Antibiotics for Lung Diseases Like Cystic Fibrosis to Be Presented at 2014 Respiratory Drug Delivery Conference

Pulmatrix Inc., a biotech company developing novel inhalation therapeutics using its proprietary iSPERSE (inhaled small particles easily respirable and emitted) technology, recently released data for its pre-clinical studies on the powdered formulation of the antibiotic levofloxacin, PUR0400. The iSPERSE formulation promises to deliver drugs in a fine dry powder form, and ensure proper and efficient dispersion within the lungs because of its unique formulation. This makes the drug more effective. This technology can be used for any drug and all possible combinations of drugs as well, as per the demand.
http://tinyurl.com/offwl25

Ataluren Phase 3 Trial Results in Nonsense Mutation Cystic Fibrosis Published in The Lancet Respiratory Medicine

PTC Therapeutics, Inc., announced that the results of a Phase 3 study of ataluren in patients with

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nonsense mutation cystic fibrosis (nmCF) demonstrated positive trends in both the primary endpoint, lung function as measured by relative change in % predicted FEV1 (forced expiratory volume in one second) and in the secondary outcome measure, rate of pulmonary exacerbations. Ataluren is a protein restoration therapy designed to enable the formation of a functioning protein in patients with genetic disorders caused by a nonsense mutation. The collective data from this trial, including retrospective and subgroup analyses support the conclusion that ataluren was active and showed clinically meaningful improvements over placebo in these trials. Safety results indicate that ataluren was generally well tolerated. Most treatment emergent adverse events were of mild (Grade 1) or moderate (Grade 2) severity, and no life-threatening adverse events were reported. Most serious adverse events reported in this study were CF pulmonary exacerbations and were considered unrelated to ataluren treatment.
http://tinyurl.com/of2okxt

Cystic Fibrosis and Diabetes Link Explained

Cystic fibrosis causes damage to the pancreas, where the blood-sugar regulating hormone insulin is produced. The mutated gene that causes cystic fibrosis also plays an important role in the release of insulin. The mutation in the cystic fibrosis gene inhibits the secretion of insulin into the blood, which means that the level is insufficient when the demands on the insulin increase, such as after a meal. The cystic fibrosis gene plays an important role in the complex chain of events that precedes the release of insulin.
http://tinyurl.com/psq73eb

Mucin Concentration Contributes to a Sticky Situation in Cystic Fibrosis

It has recently been proposed that differing concentrations of mucin within mucus layers of the CF lung contribute to decreased mucus clearance; however, it has been challenging to accurately access mucin concentration. In this study, mucin concentrations and partial osmotic pressure were greater in CF secretions compared to normal secretions. Importantly, increased mucin concentration and partial osmotic pressure promoted mucus stasis, thereby contributing to lung infection and inflammation in CF.
http://tinyurl.com/klp2bzh

TREATMENTS

Tobramycin Inhalation Powder in Cystic Fibrosis Patients: Response by Age Group. Geller DE1, Nasr SZ, Piggott S, He E, Angyalosi G, Higgins M. Respir Care. 2014 Mar;59(3):388-98.

Tobramycin powder for inhalation (TIP) is a drug-device combination designed to reduce treatment time and improve ease of use compared with tobramycin inhalation solution (TIS) in cystic fibrosis (CF) patients. However, the ability of patients to use dry powder inhalers, and the efficacy of the treatments, may vary by age. This study concludes that TIP is comparable to TIS in efficacy outcomes and safety profile but had greater patient satisfaction in all the age groups.
http://tinyurl.com/mbxd4y9

Inhaled Versus Nebulised Tobramycin: A Real-World Comparison in Adult Cystic Fibrosis (CF). Harrison MJ, McCarthy M, Fleming C, Hickey C, Shortt C, Eustace JA, Murphy DM, Plant BJ. J Cyst Fibros. 2014 May 9. pii: S1569-

1993(14)00091-5.

This real-world study compares safety, effectiveness and tolerability using tobramycin inhaled powder (TIP) versus tobramycin inhaled solution (TIS). Inhaled powder tobramycin in CF is associated with improved adherence, tolerability and decreased exacerbation rates compared to nebulised treatment in real-life practice.
http://tinyurl.com/p8qfjgl

Colistimethate Sodium Dry Powder for Inhalation: A Review of Its Use in the Treatment of Chronic Pseudomonas aeruginosa Infection in Patients with Cystic Fibrosis. Conole D, Keating GM. Drugs. 2014 Mar;74(3):377-87.

Historically, the polymyxin antibacterial colistin has been administered as intravenous or nebulized colistimethate sodium in patients with cystic fibrosis (CF) and chronic Pseudomonas aeruginosa (Pa.) infection. More recently, colistimethate sodium has been formulated as a dry powder (Colobreathe®) to be administered via a hand-held Turbospin® inhaler. Compared with nebulized colistimethate sodium, the colistimethate sodium dry powder for inhalation (DPI) formulation reduces treatment time and improves patient convenience. Colistimethate sodium DPI is generally well tolerated, with a similar adverse-event profile to that of nebulized tobramycin. Thus, colistimethate sodium DPI administered via the Turbospin® inhaler is a useful option for the treatment of chronic Pa. infection in patients with CF aged ≥6 years.
http://tinyurl.com/n75xj9j

Sinonasal Inhalation of Dornase Alfa Administered by Vibrating Aerosol to Cystic Fibrosis Patients: A Double-blind Placebo-controlled Crossover Trial. Mainz JG, Schien C, Schiller I, Schädlich K, Koitschev A, Koitschev C, Riethmüller J, Graepler-Mainka U, Wiedemann B, Beck JF. J Cyst Fibros. 2014 Mar 1. pii:S1569-1993 (14)00045-9.

Chronic rhinosinusitis significantly impairs CF patients' quality of life and overall health. The Pari-Sinus™ device delivers vibrating aerosol effectively to paranasal sinuses. After a small pilot study to assess sinonasal inhalation of dornase alfa and placebo (isotonic saline), it was found that vibrating sinonasal inhalation of dornase alfa reduces rhinosinusitis symptoms in CF.
http://tinyurl.com/ph57p9q

Nebulized Liposomal Amikacin for the Treatment of Pseudomonas Aeruginosa Infection in Cystic Fibrosis Patients. Ehsan Z, Wetzel JD, Clancy JP. J Cyst Fibros. 2014 Mar 1. pii: S1569-1993(14)00045-9.

Liposomal amikacin for inhalation (LAI) is a unique formulation of amikacin that enhances drug delivery and retention in CF airways via incorporation into neutral liposomes. Results of a recent Phase II trial suggest that LAI, with the capacity for once-daily dosing and prolonged off-drug periods, may be an attractive choice of inhaled antibiotic to manage Pseudomonas aeruginosa lung infections in CF patients. Further data from Phase III studies assessing the efficacy and safety of LAI should better elucidate its potential.
http://tinyurl.com/p75kmq9

BACTERIA

Identification of New Bacterial and Fungal Pathogens on Surveillance Bronchoscopy Prior to Sinus Surgery in Patients with Cystic Fibrosis. Kirkby S, Hayes D Jr, Ginn-Pease M, Gatz J, Wisely CE, Lind M, Elmaraghy C, Ryan-Wenger N, Sheikh SI. Pediatr Pulmonol. 2014 Apr 15.

Flexible fiberoptic bronchoscopy

Continued on page 30

CLUB CF ONLINE

The focus of Club CF is: LIVING BREATHING SUCCEEDING. Club CF wants those who have CF or are affected by the disease to see that, despite all the challenges that come along with cystic fibrosis, it is possible to live a happy and successful life.

Club CF shows how people in different age groups (20+, 30+, 40+, 50+, 60+, caregivers) are succeeding. Through Club CF, people can give hope and inspiration to those who are hesitant or nervous about what lies ahead of them.

People with CF are succeeding and making a difference in the world in high school, college, sports, careers, relationships, starting a family, post transplant, and disability. If you are one of the many people who are LIVING BREATHING SUCCEEDING, join Club CF and show the world what you have done! To learn more, please visit us online at: www.clubcysticfibrosis.com

Club CF is sponsored by The Boomer Esiason Foundation, which is committed to showing the world that people with CF are living longer and fuller lives, and by generous support from Genentech.

was performed prior to functional endoscopic sinus surgery (FESS) while under general anesthesia to collect bronchoalveolar lavage fluid (BALF) for lower respiratory tract cultures in patients with cystic fibrosis (CF). Surveillance BALF cultures obtained prior to FESS identified bacterial and fungal pathogens not previously detected by sputum or OP swab cultures in a cohort of CF patients with chronic sinus disease. Moreover, the identification of these new pathogens altered clinical management in a small number of patients.
http://tinyurl.com/nzblt66

Increasing Nontuberculous Mycobacteria Infection in Cystic Fibrosis. Ophir Bar-On, Huda Mussaffi, Meir Mei-Zahav, Dario Prais, Guy Steuer, Patrick Stafler, Shai Hananya, Hannah Blau. Journal of Cystic Fibrosis. Published online 09 June 2014.

Nontuberculous mycobacteria (NTM) are emerging infections in the CF population. Within our CF clinic, NTM incidence and prevalence have increased dramatically, associated with a severe CF genotype and phenotype. *M. abscessus*, the most prevalent NTM, caused prolonged infection despite therapy. There has been some decrease in the prevalence of NTM lung disease since 2009.
http://tinyurl.com/knutg8k

FYI

Determinants of Exercise Capacity in Cystic Fibrosis Patients with Mild-to-moderate Lung Disease. Pastré J, Prévotat A, Tardif C, Langlois C, Duhamel A, Wallaert B. BMC Pulm Med. 2014 Apr 30;14(1):74.

Adult patients with cystic fibrosis (CF) frequently have reduced exercise tolerance, which is multifactorial but mainly due to bronchial obstruction. The aim of this retrospective analysis was to determine the mechanisms responsible for exercise intolerance in patients with mild-to-moderate or severe disease. It was determined that exercise limitation in adult patients with CF is largely dependent on FEV1 in patients with severe lung disease and on the magnitude of the ventilatory response to exercise in patients with mild-to-moderate lung disease.
http://tinyurl.com/ngar6gg

Monitoring Cystic Fibrosis Lung Disease by Computed Tomography. Radiation Risk in Perspective. Kuo W1, Ciet P, Tiddens HA, Zhang W, Guillerman RP, van Straten M. Am J Respir Crit Care Med. 2014 Jun 1;189(11):1328-36.

The aim of this review is to discuss the risk of routine CT imaging in patients with CF, using current models of radiation-induced cancer, and to put this risk in perspective with other medical and nonmedical risks. The authors conclude that the risk related to routine usage of CT in clinical care is small. In addition, a life-limiting disease, such as CF, lowers the risk of radiation-induced cancer. Nonetheless, the use of CT should always be justified and the radiation dose should be kept as low as reasonably achievable.
http://tinyurl.com/l48aw9u ▲

Laura is 66 and has CF. She is a former Director of USACFA. She and her husband, Lew, live in Northville, MI.

In Memory

Michelle Marie Dixon, 23
Pawleys Island,
South Carolina
Died June 27, 2013

Nahara Mau, 56
Fremont, CA
Died April 5, 2014

Charles Barry Woodward, 60
Jacksonville, FL
Died November 24, 2013

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

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An independent, nonprofit, international organization committed to improving the quality of life of transplant recipients and their families and the families of organ and tissue donors. For information, write to: TRIO, 2100 M Street NW, #170-353, Washington, DC 20037-1233 or e-mail them at: info@trioweb.org.

American Organ Transplant Association (AOTA): Phone: 1-713-344-2402 http://aotaonline.org/default.aspx
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