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

Clinical Trials: A Vital Part of Cystic Fibrosis Treatment Advancements

By Meranda Sue

Clinical trials are a vital part of advancing treatments for CF patients. Fundraising is the genesis for new treatments because it allows researchers to create new treatments, but clinical trials are the bridge from a breakthrough in the lab to a new medicine actually being available on the open market. It is the often overlooked last step in getting a new medication approved by the FDA for the open market. As a consequence, new medications languish for extended periods because the researchers cannot recruit enough participants in the clinical trials.

As a 30-year-old with CF, I recognize the importance of clinical trials as I have been alive to see many great breakthroughs for CF. I have participated in various clinical/research trials in CF for over two decades. Some of these studies have involved mucociliary clearance, drug studies, observational studies, genetic marker studies and early intervention studies. Currently, I am participating in a study drug clinical trial and am thrilled with the ease of taking the study medication.

MERANDA SUE

The CFF drug development pipeline has never looked as promising as it does today. There are several medications with great potential being studied in CF patients. There are studies ranging from CFTR modulation which Vertex is currently leading. Vertex's Kalydeco was successfully approved after extensive clinical trials in CF patients with G551D mutation. Vertex also has VX-809+Ivacaftor (Kalydeco) and VX-661+Ivacaftor (Kalydeco) in trials for CF patients who are homozygous F508delta as well as those who are heterozygous F508delta.

Additionally, there are first-ever inhaled antibiotic therapies being

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CF ROUNDTABLE
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EDITOR'S NOTES

Welcome to winter! I hope that all have survived the Polar Vortex and were able to stay warm and healthy. What a crazy blast that cold air was. I hope that the rest of winter will be kinder to all.

We send a huge "Thank you" to all who sent donations in our "Pay It Forward" campaign. On page 10, you can see the names of the people who donated before the end of the year. Your gifts are gratefully appreciated and will be put to use in producing *CF Roundtable*.

By now, you may have read the article by **Meranda Sue** on the front page. Please consider participating in clinical trials. They are a necessary step to new meds and treatments.

We welcome two new Directors to USACFA's Board. **Lisa Cissell** and **Laura Mentch** each bring new ideas and talents to USACFA. Meet them on page 19.

Beth Sufian answers important questions about the Affordable Care Act and health insurance coverage in "Ask The Attorney" on page 4.

Then, in "Wellness" on page 20, **Julie Desch** discusses strength and how it affects our overall health. **Jennifer Hale** continues the theme of strength in "Coughing With A Smile", on page 22.

Jim Chlebda offers some more of his beautiful photos and words of wisdom in "Creative Disengagement" on page 18.

As usual, **Laura Tillman** has compiled a marvelous collection of news from various sources in "Information From The Internet". It starts on page 13.

Be sure to check out "In The Spotlight", on page 24, where **Jeanie Hanley** and **Andrea Eisenman** are featuring **Nathan Wiebe**. On page 12 we have a "Voice from the Roundtable", with **Mark Levine** talking about having one's child go off to college.

Our Focus topic in this issue is **Memory Problems**. In "Spirit Medicine", on page 6, **Isabel Stenzel Byrnes** writes of memories of the past and of her late sister, Ana. In "Speeding Past 50", on page 8, I discuss my memory glitches and how I handle them. Although Isa and I both touched on the Focus topic, I guess that most people forgot to write about their memory problems. (That's a joke, folks.)

Check out our future Focus topics in "Looking Ahead" on page 3. Perhaps there is a topic that interests you and will move you to write for this newsletter. Of course, as always, you may write about any CF-related topic that you think will be of interest to our readers. If you feel that you aren't a good enough writer, just contact us and we will be happy to help you get your thoughts down. We give priority to things that are written by adults who have CF. Then, if there is room, we may print things that are written by friends and family members of those who have CF, as long as it is of interest to adults who have CF.

Until next time, stay healthy and happy,

Kathy

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

Michelle Allen
Portland, OR
62 on September 14, 2013

Christopher Beckett
Akron, OH
47 on June 24, 2013

Bill Coon, Jr.
Pearland, TX
54 on November 1, 2013

Scott W. Kelly
Fayetteville, GA
46 on November 8, 2013

David M. Lantz
Holland, MI
43 on November 18, 2013

Paul Quinton
San Diego, CA
69 on September 17, 2013

Patricia Spadafora
Baldwin, NY
59 on December 5, 2013

Toni Marie Villines
Meriden, CT
42 on November 26, 2013

Wedding

Michelle & Gary Allen
Portland, OR
17 years on July 27, 2013

Toni Marie & Ken Villines
Meriden, CT
16 years on October 18, 2013

NEW BEGINNING

Ilana & Peter Santa
Millstone Township, NJ
Their first baby, Anthony
On September 23, 2013

LOOKING AHEAD

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

Winter (Current) 2014: Memory Problems.

Spring (May) 2014: Maintaining Mental Health. (Submissions due March 15, 2014.) Do you have any tips for maintaining an even keel on the rough seas of life? Have you found something that really works for you? Do you use medications, counseling, or something else to keep your mental balance? Please tell us how you manage.

Summer (August) 2014: Dealing With Conditions That Are Part Of CF. (Submissions due June 15, 2014.) Do you have CFRD, GERD, DIOS, CF-related arthritis, sinus disease, liver problems or any other CF-related conditions? How do you handle them?

Autumn (November) 2014: Dealing With The Death Of A Loved One With CF. (Submissions due September 15, 2014.)

ASK THE ATTORNEY

Important Information Regarding The Affordable Care Act

By Beth Sufian, JD

In the past three months readers have asked many questions about the Affordable Care Act. *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 the Affordable Care Act, accessing insurance coverage through the federal or a state Healthcare Exchange or questions about Social Security benefits .

1. When is the deadline to sign up for a health insurance policy under the Affordable Care Act?

In order for health insurance coverage to start on January 1, 2014, enrollment in the health insurance plan must be made by December 23, 2013. However, individuals can purchase a policy until March 31, 2014, on a Healthcare Exchange. Some states have set up their own Healthcare Exchanges which can be accessed online. There are 34 states that have not set up their own Healthcare Exchange and individuals who live in those states and who want to purchase health insurance coverage can go to the federal Healthcare Exchange found at www.healthcare.gov. If a person purchases a policy on January 14, 2014, the coverage will begin February 1, 2014. If a person purchases a policy on February 14, 2014, then the coverage will begin on March 1, 2014. If a person purchases a policy on March 14, 2014, then the policy will start on April 1, 2014. A policy needs to be purchased by the 15th of the month in order for coverage to begin on the 1st of the coming month. However, in December 2013, the policy can be purchased by December 23, 2013, and the policy coverage will start on January 1, 2014.

2. How do I know if an insurance policy will provide coverage for treatment at my CF Care Center?

It is easiest to contact your CF Care Center and ask them if a certain insurance company provides coverage for care at the CF Care Center. Sometimes HMO policies or other types of policies restrict coverage to certain physicians and hospitals. It is best to make sure the insurance policy provides coverage for treatment by your CF Care Center and hospital before purchasing a policy.

3. Do I have to purchase a policy on a Healthcare Exchange?

No, a person does not have to purchase a policy on a Health Care Exchange. However, if a person wants to receive a government subsidy to help purchase the insurance coverage then the policy must be purchased on a Healthcare Exchange. If a person is not interested in receiving a subsidy to purchase insurance coverage, or the person has a high income which makes him ineligible for a subsidy, the person can purchase a health insurance policy from an insurance company or an insurance broker.

> *"It is best to make sure the insurance policy provides coverage for treatment by your CF Care Center and hospital before purchasing a policy."*

BETH SUFIAN

4. If I already have a private health insurance policy do I have to purchase a policy on the Healthcare Exchange?

No. If a person already has insurance coverage from their employer, their parent's employers, their spouse's employer, Medicare, Medicaid, Tricare, or an insurance policy that provides comprehensive coverage, then the person does not need to purchase another policy on the Healthcare Exchange.

5. Can an insurance company charge me more for my health insurance coverage because I have CF?

According to the Affordable Care Act, starting on January 1, 2014, an insurance company is prohibited from refusing coverage to a person due to a

pre-existing condition like CF. The insurance company cannot increase the premium for the policy because a person has CF or any other pre-existing medical condition.

6. If I lose my current coverage after March 31, 2014 can I go on the Healthcare Exchange and purchase a policy?

Yes. If a person loses coverage after March 31, 2014, the person can enroll in a policy offered through a Healthcare Exchange or purchase a policy through an insurance broker or insurance company.

7. I am 21 years old. Can I continue on my parent's health insurance policy even if I am married?

Yes. According to the Affordable Care Act, an adult child who is under the age of 26 can continue on their parent's policy until the adult child reaches the age of 26 (see #8 for an exception that expired on December 31, 2013). The adult child does not have to prove she is a student or that she is unable to work.

Once the adult child turns 26 she can continue on her parent's insurance policy in most states if she is incapable of self-support. Continuation of coverage will require that the adult child's physician complete a form provided by the insurance company that states that the adult child is incapable of self-support. Different states may have other requirements that must be met in order to continue coverage under a parent's insurance coverage once the adult child reaches the age of 26.

8. I am 24 years old and I was told that I could not continue on my parent's insurance policy if my employer offered health insurance coverage to me as an employee. Is this true?

Until December 31, 2013, an adult child who was working and whose employer provides health insurance was required to enroll in the employer sponsored health insurance policy. The adult child under the age of 26 is not allowed to continue on the parent's policy if the adult child has an employer who provides health insurance.

However, on January 1, 2014, even if an adult child has an employer who provides health insurance coverage, he can choose to stay enrolled under his parent's health insurance policy as long as he is under the age of 26. This can be helpful if the parent's insurance policy provides better coverage than the health insurance policy offered by the adult child's employer.

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 free 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 over 32,000 calls from people with CF, their family members and their healthcare providers since 1998. ▲

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.

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 individual's 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.

SPIRIT MEDICINE

The Spirit of the Past

By Isabel Stenzel Byrnes

"Look to this day for it is life. In its brief course lie all the realities and truths of existence. The joy of growth, the glory of action, the splendor of beauty… today well-lived makes every yesterday a memory of happiness and every tomorrow a vision of hope. Look well, therefore, to this day."

--Ancient Sanskrit proverb

In this issue of Spirit Medicine, we highlight the topic of memory problems. Note the word 'problems'. When we can't remember something that happened, it bothers us. So I will muse about spiritual aspects of the past. This little four-letter word, 'past,' informs who we are; it is responsible for our personal narratives. Stories from the past make up the bulk of our social conversations. Is it important to remember our past, and if so, why? Does *how* we remember our past matter? Also, does having cystic fibrosis (CF) affect our view of the past?

The other day, I was buying an airplane ticket online and I found myself scrolling *waaaaay* down for my birth year. Each of those years was filled with memories, milestones and the people who crossed my path. As a person with CF, I felt grateful and proud for all that time. The older we get, our suitcases of memories just gets heavier. Our brains are not capable of remembering every single moment. Fortunately, or unfortunately, we usually remember emotionally charged events best.

With each life experience comes a collection of emotional and psychological reactions, and responding well to those reactions leads to growth. In essence, our maturity depends on how we've dealt with the past. Dealing with traumatic events in the past is one of life's greatest charges. Rather than being immobilized by hardship that has happened, Robert Emmons writes, "Contrasting the present with negative times in the past can make us feel happier (or at least less unhappy) and enhance our overall sense of well-being. This opens the door to coping gratefully."

> *"With each life experience comes a collection of emotional and psychological reactions, and responding to those reactions well leads to growth."*

ISA STENZEL BYRNES

The past exists in our minds, distinctly different from our present world. So the great opportunity here is that we have the ability to change our emotional reactions to memories. What might have been seen as a horrible, meaningless event may later bring up new insight and perspective. We can re-construct the past, in essence, in our minds. We can use positive psychology and tell ourselves things like, "I'm not the only one who endured this. I've learned what I'm made of. If this bad thing didn't happen, some of these other good things wouldn't have happened either."

And also, when we do remember good times, our memories carry the spiritual power to bring us back to a time and place that once made us happy. For example, neuroscience tells us that if we use our minds and just *imagine* being held by a loved one, then our brain chemistry can actually release oxytocin, the attachment hormone. It's as if we are bringing the past back to our bodies, right here, right now.

Having (CF) inevitably informs our past. Our past interactions with doctors will impact our current trust of doctors. If we had a negative experience with a medication, we will always remember not to use that medication. We are constantly monitoring what worked and didn't work to address our current treatment needs.

CF is not a part of all my memories. We humans have selective memory: sometimes consciously and

unconsciously we choose what we remember. For example, I don't remember much about coughing as a kid. I know I coughed; my mother told me I coughed all the time, but I don't remember it at all.

However, CF will often bring up complex feelings when we reminisce about the past. It is a mixed blessing that this disease is progressive, as most of us have a past with relative health. We might look back at the time when we were on the high school track team, and now we find it hard to walk a mile. Some of us may *yearn* for a time when we were healthier; when we could work or travel or keep up with friends. Some of us may remember taking our health for granted, and doing things to our bodies that we now regret, such as skipping treatments. Those memories might bring up feelings of sadness, guilt, or despair.

Some of us who received lung transplants may remember a time when we were much sicker than we are now. Reflecting on the past, and how far we've come, will bring up feelings of joy, gratitude, relief, confidence. Sharing our past, and the contrast in our health, is what makes this transplant story of suffering and redemption so miraculous.

Many of you know that I lost my twin sister, best friend and soul mate, Ana, this past September. Like many who are grieving, I have to sort through Ana's things. Stuff that was once so present is now past for Ana. I found letters from friends, journals, yearbooks, and mountains of travel scrapbooks. Ana, like me, was obsessed with documenting in the present, so she could cherish her past; it proved that she existed. My sister's belongings from the past carry her energy. These things comfort me, as they are evidence Ana lived a full life.

As part of the grieving process, I wish I could rewind and return to a time when Ana was with me. It would be really easy for me now to live in the past. Being with Ana – whether we were hospitalized together or caught up in a sisterly catfight – were the best times of my life. While I want to honor the time I had with Ana, if I dwell too much in what once was, I'd miss out on the opportunities and relationships that are present today. And grabbing on too hard to what I once had raises the reality of another four-letter word: gone. It hurts.

My past with Ana has made me who I am today. So now I have her influence to carry me forward into my future. Her memories–our shared memories–shape my psyche, and give me spiritual strength to move forward. But I already know I cannot remember every single moment of our lives together.

So what are memories, really? I like to believe that memories are bundles of spiritual energy that are created in the moment, but do not actually disappear when we "forget". They simply dissipate into the universe, sometimes to be captured by other people who can share the memory or remind of us what happened. When Ana's friends share a memory with me, it feels bigger and stronger than my remembering alone. But sometimes the energy is just held, out there, somewhere. It joins a collective consciousness. The past never dies.

With this belief, is a memory problem really a "problem"? I am reassured when I will forget, that it's okay. It doesn't matter. God remembers. God doesn't forget. This is tremendously comforting. We are not the only keepers of our memories.

I have often heard that right before a person dies, he or she will "see" glimpses of every moment in the past. I am comforted by a spiritual perspective of time. In her book, "Dying to Be Me," Anita Moorjani undergoes a powerful near-death experience. She writes that when she crossed over, time had no meaning anymore; that she reviewed images from her life and basically remembered everything and everyone all at once. I'd like to believe that. That it all is connected; that it is all here and now, past present and future. In a blink of an eye, my life will be over and I will be reunited with Ana. Of course, I still have a strong will to live. Whether I live 5, 10 years, 15 years more, it will seem like a split second. And indeed, in the meantime, I will cherish the past memories that I can remember, let go of the ones I can't, and be mindful of the present. Deeply, wholly focused on the present. And I will be grateful. At times of forgetfulness, may you all be blessed with similar comfort. ▲

Isabel Stenzel Byrnes is 42 and has CF. She lives in Redwood City, CA with her husband, Andrew. You may contact her at: isabear27@hotmail.com.

In Memory

Harold (Hal) Soloff, 82
Norwich, CT
Died on September 23, 2013

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

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

SPEEDING PAST 50...

Memories

By Kathy Russell

Life is good. Sounds simple, doesn't it? Sometimes it is, but other times it isn't. In October I had bilateral mastectomies because of cancer. I chose to have no reconstructive surgeries and I am fine with that. My aching back doesn't miss my old front. I don't have a problem with looking like a bowling pin and my husband still loves me, so all is good. …and life goes on.

We have gone from the most lovely summer I can remember, through a pleasant autumn and into a sudden arctic blast of winter. I am so grateful for our efficient woodstove. Even when the east wind blows with a vengeance out of the Columbia River Gorge, we stay cozy and comfy. If it is too nasty outside, I don't have to go out for anything. How spoiled am I? I hope you are warm and cozy, too.

I noticed that the Focus topic for this issue is: "Memory Problems". I have a lot of experience with that. It seems that as I get older my memory gets more full of holes. I attribute some of that to my many bouts with general anesthesia. The rest of it probably is due to my advanced age and all the medicines I take and have taken. I do try to limit the number of meds I take, but it seems that there always is something more that I have to take.

Some memory loss is inevitable, but other losses can be mitigated. I love to play memory games. I feel that they help me to keep some of my mental acuity. Making my brain work is good for it, I think. Each challenge helps to sharpen my wits.

I live in Oregon and we have 36 counties. I like to list the counties in alphabetical order as a test of my memory. I usually can get at least 35. Not bad for an old broad. I do the same thing with the names of the states and then with their capitals. I think that such exercises help to keep my memory working.

I am short on physical strength and energy so I make up for it with mental strength and stamina. I love to take tests, even if they are on subjects about which I know nothing. I enjoy trying to figure out the answers from what the questions are asking. I usually am able to score at least 75% on unfamiliar subjects. If I know something about the subject, I expect to score at least 85%. If I start to get lower scores, then I know it is time to work on my thinking abilities.

I enjoy crossword puzzles. The puzzles that are in the NY Times, LA Times or London Times are interesting and challenging. I try to do them quickly and without any references for help. I sometimes fail, miserably. Then I go back and try to learn the items that I missed. All in all, it is good exercise for my brain.

> *"I tend to let unpleasant memories fade. I see no advantage to remembering things that make me sad, angry or unhappy."*

KATHY RUSSELL

Since I do not write as much as I used to, I find that my spelling skills get rusty. I do little spelling bees with myself. I will think of a word, try to spell it and then look it up to see how I did. Boy, do I miss badly at times. Of course, it is a long time since I was in school, so that might be one reason that my skills are not as good as they should be.

Getting adequate rest is essential to good memory. If I am tired, my brain is just mushy. I know that the answers are in there, but I can't get them to come out. It is as if my file cabinet of a brain is stuck shut. If I have enough rest, just like magic, my memory works.

I tend to let unpleasant memories fade. I see no advantage to remembering things that make me sad, angry or unhappy. Of course there are some memories which make me sad but that are important to remember. I love to remember times with friends who

have since died. Even though the fact of their death may sadden me, remembering the good times that we shared makes me happy. I like to be happy, so I work at remembering the happy times of my life. It works for me.

I am able to remember specific incidents from my early life. The earliest memory that is strong in my mind was the death of my first nephew. He was only ten weeks old and died of SIDS. (Yes, it was SIDS, not CF. He had been ten weeks premature and his lungs were not developed.) Although I was only two-and-a-half, I remember all of the goings-on very clearly. I was excited about becoming an aunt. I knew what that meant and it was cool. My sister had another baby a year later, so I got to be an aunty again. Yes, I remember when he was born just as I remember the subsequent births of many nieces and nephews.

At times, I try to remember anything about a specific year. Some years just do not stand out. Other years make it easy for me and I am able to come up with many happenings. I use clues such as photograph albums, scrapbooks, presidential elections, hit movies etc. to come up with something. It is amazing what one can remember with a little help.

So I have talked about long-term memory; now let's discuss short-term memory. That can be a problem. I can read something in the newspaper, put down the paper, get up to walk into Paul's office to tell him what I read and have forgotten it before I get there! I really hate doing that. I try to think of something that can lock it into my memory. It may be as simple as crossing two fingers or as weird as repeating the headline all the way to Paul's desk. Once I get there, if I have remembered what I was going to say, I still may forget particulars of the story. Aaarrrggghhh!!! Old age hits again.

I try to concentrate on what someone is telling me so that I won't be caught with my mouth hanging open, catching flies, because I have zoned out. Zoning out is a real problem for me. I forget to pay attention (sometimes) and am caught looking stupid. If I have done this to you, I apologize. It seems to be a symptom of aging and being tired.

> *"All the lists in the world won't do much good, if you don't look at them. Maybe I need to make a list of my lists and where they are."*

Another symptom of aging is the difficulty that I sometimes have with remembering a person's name. I can look at someone and know that I know them, but their name just will not come up to the front of my mind. If only there were some way for me to look in the mirror and read what seems to be "on the tip of my tongue". Oh, if only…

I have tried some of the recommended exercises to sharpen my memory of people's names, but it never has seemed to work for me. I'll remember the image I selected, but the name of the person still isn't there. Many times I just tell the people that I have a hole in my memory and ask them to tell me their names again. Fortunately, most of our friends are in the same boat, as far as memory goes, and they understand.

At least I seem to be able to remember medical appointments and newsletter deadlines. I'd be in a world of hurt if I couldn't keep those straight. A good calendar, with large spaces for the days, helps me with those items. As soon as I make an appointment, I write it on the correct space of the kitchen calendar. That is where both of us look to see if we have any commitments on any given day. It helps both of us.

Since the deadlines for this newsletter are the same every year, they are pretty easy to remember. Copy needs to be here by the 15th of March, June, September and December. Then we can work on getting it ready for the printers. I need to remember to leave enough room in my schedule for editing and getting the copy to Andrea for the layout. She needs to have enough time to work on it at her leisure. As long as we all remember to leave adequate time, all goes smoothly.

I make lists to help me remember things I need to do or items I need to buy. Then I just need to remember to look at my lists. I am famous for forgetting to take my grocery list with me to the store. All the lists in the world won't do much good, if you don't look at them. Maybe I need to make a list of my lists and where they are. Perhaps that would help, but only if I remember where it is and look at it.

At any rate, remember to keep working on your memory. A working memory can make life much better.

Until next time, stay healthy and happy. ▲

Kathy is 69 and has CF. She is Managing Editor of CF Roundtable. She and her husband, Paul, live in Gresham, OR. You may contact her at: krussell@usacfa.org.

PAY IT FORWARD

On behalf of the board of directors of USACFA, I'd like to thank everyone who donated to our first fundraising campaign since *CF Roundtable* has become available free of charge. While there is no cost to our readers, we still have printing, mailing, website maintenance, and other related expenses. In order to continue to provide our newsletter for free, your generous support is greatly needed and appreciated. In all honesty, we were overwhelmed by your contributions! You exceeded our expectations and have provided enough funds to publish our next issue of *CF Roundtable*. We thank you for thinking so highly of us and hope that 2014 brings you a life lived well, regardless of whatever comes your way.

Below you will find a list of our terrific donors who participated in paying it forward!

Laura Tillman
President

Colleen & Scott Adamson
Paul Albert (in memory of my organ donor)
Michelle Allen
Derwin & Blanche Ball (in memory of Marsha Ball-Waldo)
K. Gerald & Ruby Balls (in memory of Greg Baxter)
Cheryl T. Baum
Christopher Beckett
Joan Finnegan Brooks
Ingrid Chartrand (in memory of Sonya Chartrand)
Peggy Clark (in memory of Keven Kent)
Monica Coburn (Just happy to be alive.)
Toddy Coon (in memory of Diane Farley Coon)
Toddy Coon (in honor of Bill Coon, Jr.)
Edward & Elaine Corr
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Kitty Fortier (in memory of David Paul Thompson)
Judy Greenberg (in honor of Ilana Greenberg Santa)
Douglas & Barbara Gonzalez (in honor of Steven J. Gonzalez)
Steven Gonzalez (in memory of Douglas A. Gonzalez)
Patty Hardesty (in honor of Vicki Zacher)
Dr. & Mrs. Douglas Holsclaw (in honor of Dr. Paul Quinton)
Anne Marie Jewell (in memory of Debbie Lynn Ajini)
Jennifer Jessie & John Dungan (in honor of Cynthia Dunafon & Steven Holloway)
Theodore Kowalski & Philip Devine (in memory of Joseph Kowalski)
Dave & Elaine Lantz (in honor of our son, David M. Lantz)
Betty Lefer
Hedy McLaughlin
Robert McLaughlin
NJ State Organization of CF
Kim & Joe Nunnari
Stephanie Rath (Go USACFA!!)
Jeannine Ricci
Kathy Russell (in memory of Anabel Stenzel)
Kathy Russell (in honor of the USACFA, Inc. Board of Directors)
Maryanne Sanchez (in memory of Sherry DiTursi)
Stephen Scheu
Jane Schnackenberg
Phyllis Sewell
Janice Siegel (Thank you for all you do!)
Patricia Spadafora
Penny Stroud
Frank & Judybeth Smith (in honor of our grandson, Michael J. Kozlowski)
H. R. & Dorothy Stewart
Robert T. Stone MD
Barbara J. Tews
Susan Tickell (in honor of the Desch family)
Laura & Lew Tillman (in honor of all those with CF)
Debra Tumillo
Maria Valdes-Domingoes
Nancy Marcuz Wech (in memory of Lauren Melissa Kelly)
Nancy Marcuz Wech (in honor of Scott W. Kelly)
C. Barry Woodward
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Mailbox

Happy Holidays and Merry Christmas to all associated with publishing the wonderful *CF Roundtable*. Many thanks for your hard work.

Toddy Coon
Tomball, TX

Enclosed is a small donation for your newsletter. As a mother of a 39-year-old, I enjoy it very much and share it with my daughter.

Connie Flowers
Middleton, OH

Great work!

Jim Yankaskas
Chapel Hill, NC

So much information. Many thanks.

Edward & Elaine Corr
Taunton, MA

We love this magazine for all your terrific work!

Cheryl T. Baum
Lake Zurich, IL

I am 48 have CF and am a breast cancer survivor. I enjoy the newsletter very much.

Maria Valdes-Domingoes
St. Augustine, FL

Hi there,
I enjoy reading your newsletter and thank you for producing it.

Two quick questions:

1. The articles are so well written that I'm wondering - are your contributors paid for their submissions - by way of fee or honorariums - or are the entries entirely voluntary?

2. Also, once submissions are made, does anyone edit them or proof them for grammar - or is that the responsibility of the person submitting the article?

Thank you,
Quin Tran
via e-mail

Editor's response: Thank you for your kind words. Our authors are volunteers and receive no compensation. Most are adults who have CF and have something that they want to say.

We do edit for clarity and make some changes for grammar. We try to leave things in the voice of the author, as much as possible.

Also, we will assist anyone who wants to write but feels that he/she is not really an accomplished writer.

Thank you very much for making this [*CF Roundtable*] online. I enjoyed being able to read the newsletter and learn more of the success and perspectives of adults living with CF.

Best regards,
Leslie Hazle, MS, RN, CPN
Director of Patient Resources
CFF

My wife, Kathleen Harris, and I have been avid readers and supporters of *CF Roundtable* since at least 2005 so we were very pleased that you agreed to publish her two "milestones" – her 70th birthday on May 17 and our 50th wedding anniversary on June 22. These were two big items on her modest bucket list. Unfortunately, Kathleen never saw the last edition of *CF Roundtable*. She passed away on June 27 from ovarian cancer, and kidney failure – both, at least in part, the result of seven years of immune suppressant medications. Even so, she said she wouldn't hesitate to do it again.

Kathleen was tough – boy was she tough! She coped emotionally and physically with the effects of CF and the resulting lung transplant for more than 30 years of our marriage. She rarely complained, but often would remark at how lucky she was and what a good life she had. Her family meant everything to her and she read articles in *CF Roundtable* with interest and concern about the many people who have CF who could never have children, but we had two wonderful daughters and three grandsons that really did light up her life. Kathleen was smart, though she never recognized that because of her deep sense of humility. She was sometimes blunt, in the way of her Irish heritage, but never hurtful. She was always patient, loving and kind. She had a prayer list as long as your arm and prayed hard for those on it every day. Kathleen was patient, especially in suffering the indignities of an illeostomy and a nephrostomy tube in each of her kidneys. She had a special affinity for the homeless. When she was able, she served meals at a soup kitchen in Washington DC and she cooked meals for them once a month with others in our church.

Above all she treated every person she met with the same degree of dignity. This was especially evident in

Continued on page 31

Making the Move: Moving My Step-Daughter into College

By Mark A. Levine

Last month we moved our daughter, Brooke, into college. It was quite emotional and very exciting. It was the last stage of a roller-coaster ride of a process; from campus tours to standardized testing to essays and applications. Then there was waiting to hear from schools, discussions on which one to attend, deciding on a roommate, packing and moving in. Attending classes, studying and the rest of what college life is about is all up to Brooke and it has been fun listening to her updates even though we are just a few weeks into the school year.

My wife and I know what this big change means to the family. Our daughter has moved out of the house. She will be back for Thanksgiving and, perhaps, over the summer if she does not get a job or internship or out of town boyfriend. Our home will always be her home. But as for her home being our home, that time has passed. This was a milestone of an event in which we just participated first hand, front row.

Although it was over 25 years since I started college, I remember it like it was yesterday. Quite frankly, it is hard to imagine that it was so long ago. I have to confess that my life perspective has changed considerably in these last 25 years and sending a child to college has been a great reason to reflect.

I attended Lehigh University in Bethlehem, Pennsylvania. The school is beautifully set into the side of a mountain. My residence hall was at the top of the mountain with a foreboding staircase leading up to it, as if the mountain was not enough of an obstacle already. During my first full week on campus I remember wondering if this was a good choice of a school for me. At 18 years old, I was healthy and did not have CF complications. But what happened, I remember asking myself, if I did have issues? That staircase leading up to my room might be more than I could handle.

It turned out that neither the mountain nor the staircase ever beat me. I wound up living in that area of campus for four years and loving it. After graduation, I had similar thoughts when moving into my first apartment in Michigan. Located on the third floor in a building without an elevator, I questioned my decision, hoping that I would not get too sick to be able to make the trek up to my room. When I moved out three years later to relocate to Indiana, I realized that my third floor choice had been a non-issue.

If you had asked me back then about starting a family, being a step-dad and holding a full time job for twenty years, I probably would have laughed. It was not that I was a pessimist... quite the opposite in fact. But I have always tried to be a realist and those thoughts and questions at that stage in my life seemed to be fair.

The times, however, have changed. I have stopped questioning and started doing. It is not to say that I have not had setbacks or heartaches or made mistakes. I have had my share of bad days and my share of weeks on IV antibiotics. My younger brother, David, died 20 years ago after his battle with CF without grad-

Continued on page 14

JOELLE, BROOKE AND MARK LEVINE AT NORTHWESTERN UNIVERSITY.

SUE *continued from page 1*

studied in CF patients. For example, Savara Pharma's (www.aerovanc.com) lead product, inhaled dry powder Aero Vanc, is being studied in CF patients who culture MRSA (methicillin resistant staphylococcus aureus). Aero Vanc could be a huge advancement in the treatment of MRSA CF lung infections. Currently, there are no commercially available inhaled antibiotics for MRSA. Subsequently, those with CF and MRSA have to take very strong oral and IV antibiotics for every exacerbation. For those with Pseudomonas, inhaled Levofloxacin and Arikace are also in development in clinical trials. These new inhaled antibiotic therapies have potential in lessening the bacterial burden in CF patients' airways, thus, leading to reduced airway infections and subsequent irreversible lung damage.

Participating in clinical trials is not as challenging as one may think. Patients are reimbursed for study participation, travel (mileage), parking, food consumed during study visits and sometimes overnight stays at a hotel, depending upon distance traveled by the patient. The study sponsors do their best to make study participation less inconvenient to CF patients and their families.

Great sources for online searches of CF clinical trials are www.clinicaltrials.gov, www.cff.org drug pipeline, and company websites who list study drugs in trial for CF.

Best of luck to you in your efforts to participate in CF research. Just remember to always consult your CF doctor prior to screening or joining any clinical trials. ▲

Meranda is 30 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Information from the Internet...

Compiled by Laura Tillman

PRESS RELEASES

Nitric Oxide Designated Orphan Drug for Cystic Fibrosis

Novoteris announced that the FDA has granted Orphan Drug Designation for inhaled nitric oxide gas for the treatment of cystic fibrosis. Nitric oxide has shown to be an effective antimicrobial agent against broad spectrum of microbes, including drug resistant strains. This orphan drug designation was granted based on its pilot trial that demonstrated > 15% increase in lung function following two weeks of therapy with inhaled nitric oxide gas.
http://tinyurl.com/m3h92zd

High Rates of Bowel Cancer in Cystic Fibrosis

Patients with cystic fibrosis (CF) may be up to ten times more likely to develop colorectal cancer. CF patients also had a threefold higher risk of adenomas and a sevenfold higher risk of advanced adenomas.
http://tinyurl.com/l7gudzb

The Eye in Cystic Fibrosis

Cystic fibrosis is the most common lethal autosomal disorder in Caucasian populations. It is characterized by a variable degree of pulmonary infections, pancreatic enzyme insufficiency and premature death. Ocular complications in CF range from abnormal tear volume to impaired dark adaptation. With improvements in CF life expectancy, ocular complications are of greater relevance to the optometrist.
http://tinyurl.com/mld75rp

My Dishwasher Is Trying to Kill Me: Extreme Conditions Suit Pathogenic Fungus

The article focuses on the occurrence of potentially pathogenic fungal flora located in dishwashers. 62% of the dishwashers contained fungi on the rubber band in the door, 56% of which accommodated the polyextremotolerant black yeasts *Exophiala dermatitidis* and *E. phaeomuriformis*. Both *Exophiala* species showed remarkable tolerance to heat, high salt concentrations, aggressive detergents, and to both acid and alkaline water. This is a combination of extreme properties not previously observed in fungi. *Exophiala dermatitidis* is rarely isolated from nature, but is frequently encountered as an agent of human disease, both in compromised and healthy people. It is also known to be involved in pulmonary colonization of patients with cystic fibrosis. The discovery of this widespread presence of extremophilic fungi in common household appliances suggests that these organisms have embarked on an extraordinary evolutionary process that could pose a significant risk to human health in the future.
http://tinyurl.com/ohrvlyv

TREATMENTS

Early Treatment with Inhaled Antibiotics Postpones Next Occurrence of Achromobacter in Cystic Fibrosis. M. Wang, W. Ridderberg, C.R. Hansen, N. Høiby,

Continued on page 21

LEVINE *continued from page 12*

uating from college or getting the opportunity to start a family. However, at 43 years old, my outlook on the future is nothing but optimistic.

I feel good that I have two college degrees, hold a full time job, have a wife and family and stay active working out and playing tennis. These things did not come because I spent my life second guessing myself. I know that there are some things that are beyond my control, but I also know that things happen to do those who do. You cannot plan everything out and sometimes, when you are in the middle of something special that you always thought could happen but was not really ever in your playbook to happen, it hits you.

I have to admit that way back in mind, I always thought I would have a family, but I really did not know how I was going to pull it off. I knew that I could not have kids of my own, but I still knew a family was in my future. I did not plan to meet a woman who had kids from a previous marriage but, for me, it was a formula for success.

> "If you had asked me back then about starting a family, being a step-dad and holding a full time job for twenty years, I probably would have laughed."

Looking back, I got lucky. When I met my wife, her kids were six and four. They were eight and six when we got married and I have since considered them my own. Being a step-dad has been nothing short of awesome. From daddy-daughter dances in the third grade, to cub scouts with my son, to sports with both kids and helping them with homework, teaching the kids how to ride bikes and later how to drive, the rewards have been never ending.

Did I ever think that I would have the honor and privilege of moving a daughter into college? Let me put it this way… I never thought that it would not have happened and I have never let anyone tell me that it could not happen; so now that it has happened, I am humbled, but not surprised.

Life is one of those things. It keeps on going, until it doesn't. I continue moving forward knowing that if I overstep my bounds, I can always readjust. Failing to live is not an option. You never know what lies in front of you.

My son graduates high school in two years. I will love every minute between now and then, but I know that another move-in day is right around the corner.

Mark is 43 and has CF. He is a Director of USACFA. His contact information is on page 2.

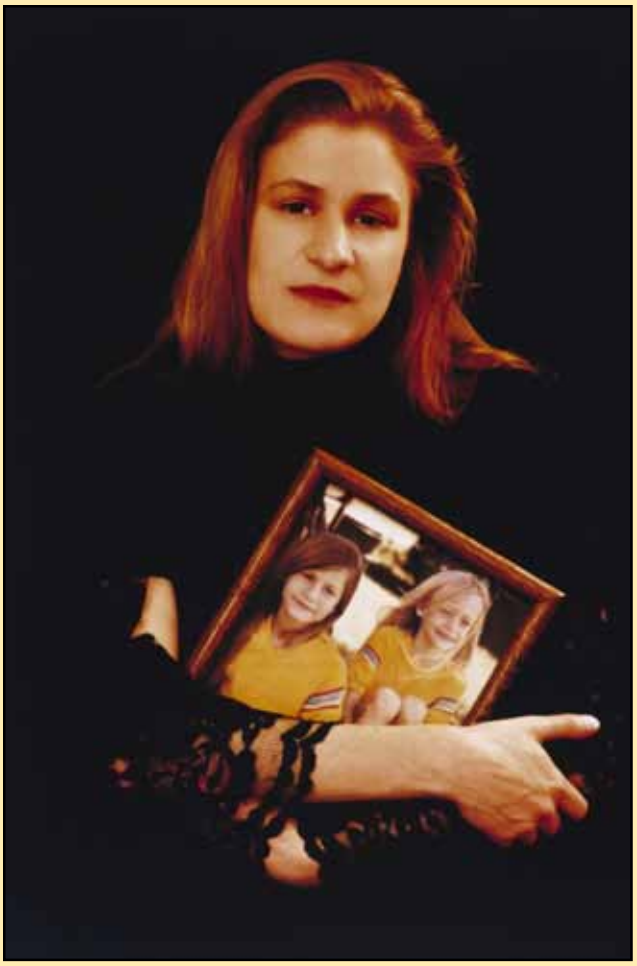


PHOTO BY DAVID MYERS

Miranda

All I have are memories and photographs
Of you so sweet yet obstinate.
Naked and rebellious
Wiser than your years
My little sister, my friend, my love
If only you hadn't died
We would be together happy and free
Sharing in our survival and pain
Only I share this with my husband now.
Come and meet my husband and my friends!
What kind of woman would you be today?
You've taught me everything I know
About independence and laughter.
Without you I am not complete
I have never forgotten you,
I will always love you.
Here on Earth my life goes on
Until Cystic Fibrosis claims my life, too.

-K. Bischoff-Howell, 1998

"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

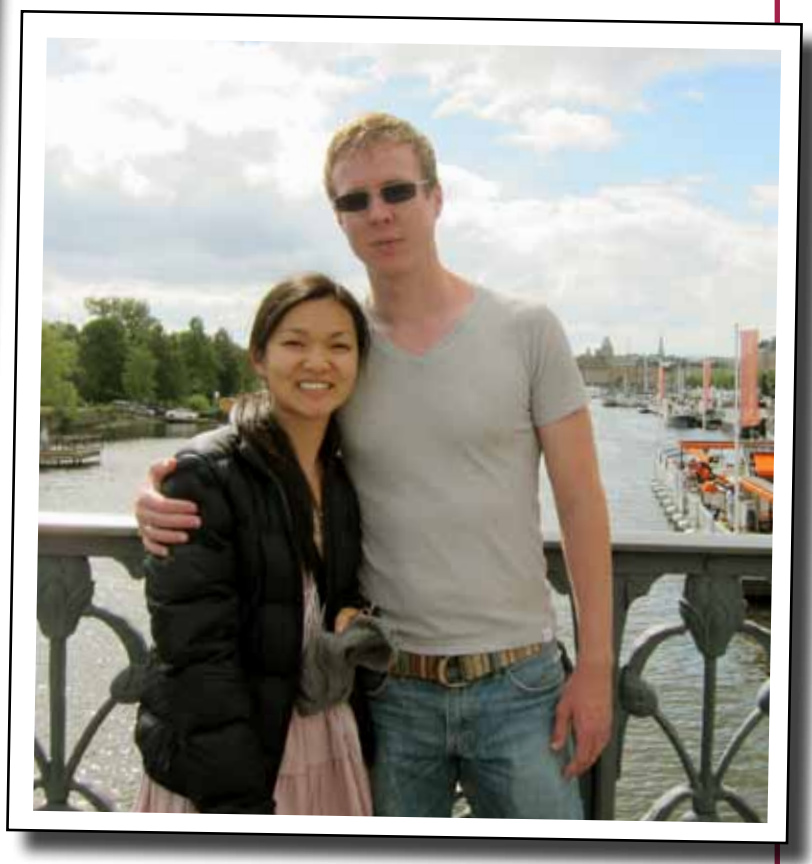
FROM OUR FAMILY PHOTO ALBUM...

"THERE IT IS! THE POT OF GOLD." LAURA MENTCH WITH HER FOUR-YEAR-OLD GRANDSON, ARLO, IN VIRGIN ISLANDS NATIONAL PARK.

PAUL AND KATHY RUSSELL IN THEIR YARD.

MARK LEVINE WITH WIFE, JOELLE, AND CHILDREN, BROOKE (18) AND ADAM (16), IN THEIR BACKYARD.

MARK AND JENNIFER HALE.



MOLLY AND NATHAN WIEBE IN STOCKHOLM, SWEDEN.

CREATIVE DISENGAGEMENT

The Ethereal Nature of the Jimson Weed

By Jim Chlebda

There's something about Jimson weed *(Datura)* that I've always been attracted to. The plant itself is *very* psychoactive—it is nothing to fool with as far as ingesting it is concerned—it is a powerful, potentially lethal plant. The original southwest desert inhabitants had ceremonial use for it, but not many of them are around today to explain particulars for its safe useage.

Jimson leaves have a very strong, obnoxious aroma. A built-in early warning system for its harmful aspects, perhaps? What is truly captivating though, are its large flowers. They are beautiful and have a light, delicate scent. But what I find most appealing is the way they capture and hold sunlight, *especially* when I've got that digital camera in hand to experiment with.

Some Jimsons can grow to large-sized shrubs, with many satiny blossoms opening almost in unison. Often the flowers have soft lavender-violet tones irradiating the rims of those creamy-white cups, which can measure five inches across. Under a full moon they're also quite striking, with a soft, pearly opalescence. I've seen cottontail rabbits nibble the leafage down to stumps; that could explain their often goofy behavior. I've isolated new plants in chicken wire cages to discourage those rascally rabbits! The bee, in the photo, I shot early on a different morning—it had spent a chilled night down inside that flower and was struggling its way up the stamens to reach what my camera also sought: that warm glow of those first rays of the sun. ▲

Jim, 56, has CF. He resides on the edge of Joshua Tree National Park in California and can be contacted through: back40publishing.com.

Meet Lisa Cissell

I am happy to be a new director on the board of USACFA!

I was diagnosed with CF at the age of 25. I am 50 now, so for half of my life, I was blissfully unaware.

I am the oldest of seven children; all girls except for one brother. Two of my sisters also were born with CF. My sister Mindy passed away in 1999 while waiting for a lung transplant. My sister Cara is currently on the waiting list at Jewish Hospital in Louisville, KY.

I live in Bardstown, which is in central Kentucky, about 25 miles from where I grew up.

I earned a Bachelor's Degree in Agriculture from Eastern Kentucky University. Currently, I work for the USDA Natural Resources Conservation Service. I have been there for 26 years.

Around the age of 40, my health began to decline and I reached the point of needing a lung transplant. In April 2011, I received that gift at Barnes-Jewish Hospital in St. Louis and have been going strong ever since.

I share my home with a cutie Shih-Tzu named Sherman and a fat, orange tabby cat named Copper.

I love to travel any chance I get. I love anything horses and I love sharing laughs with my family and friends.

You may contact Lisa at: lcissell@usacfa.org▲

LISA CISSELL

Meet Laura Mentch

I was diagnosed (finally!) with CF in 2003, the month I turned 50. After a lifetime of health issues I am grateful to be able to be in the care of health care providers who know how to help me.

While looking for information about living with cystic fibrosis, I found *CF Roundtable* and a connection with other adults with CF. Reading the experiences of others has given me so much help and encouragement.

My work has been in sexual health education and I have been fortunate to work with people of all ages in community and school settings. I am happy to bring my experience to the CF community and provide education about sexual health and adult diagnosis of cystic fibrosis.

Of the five children in my family, I am the youngest and the only one with CF. The older kids used to tell me that Mom and Dad ran out of the good genes before I was born!

Michael Brody and I have been married since 1976. We started our journey together in Massachusetts and now live in Bozeman, Montana. He is a professor of education at Montana State University. We are the parents of Adam, Luke and Cara and grandparents to Arlo and Teddy. I feel fortunate to have a family.

With gratitude for all of the adults who have helped me, I join the dedicated Board of Directors who bring you *CF Roundtable*, which is full of valuable information and resources. Please contact me by email at: lmentch@usacfa.org. ▲

LAURA MENTCH

WELLNESS

Strong Is the New Sexy

By Julie Desch

As I get older, I find myself frequently wondering how it is that I continue to do so well with my original lungs. I'm 53, and my lung function has remained fairly stable for many, many years. I'm actually quite superstitious about this subject and am almost afraid to write about it, lest somehow this causes things to change drastically as soon as this hits the airwaves. But the reason I've decided to discuss it is I happen to ***know*** that there is at least one factor that has greatly contributed to my health and has been totally under my control.

I'm under no illusion that I am in absolute control of everything CF related...I know that my DNA must contain some serious modifier genes working for me, for instance. I also know that, unlike my older siblings who didn't do as well with their CF, my parents had stopped smoking by the time I rolled around, and this most definitely helped my little lungs when I was a wee thing. These things are totally out of my control, and have certainly contributed to my run of good fortune. But there are also a few things that I think I can (and should) take credit for and I feel like I almost have a responsibility to share them so that others will benefit as well.

Nobody forced me to get strong. It was entirely my idea, and an odd one at the time. It didn't just come to me through good genetics, either. It took consistent, patient, diligent ***hard*** work on my part. It was 1980, and the big craze was to do "cardio" to get fit. I drank the jogging Kool-Aid and ran miles and miles to get my cardio in.

I will admit that my motivation to jog had absolutely *nothing* to do with CF back then; it had everything to do with vanity. I wanted to be slim...if not downright skinny. I absolutely hated running. It was hard to breathe and I found nothing about it enjoyable except the feeling that I had when it was over for the day! That feeling kept me hooked, though, and I kept at it.

Of course, this was an awesome way to treat my CF back then, and I'm sure it played a huge part my not requiring IVs until my mid-30s. I did nothing else in terms of respiratory care...this was before the days of the Vest or flutter. Pulmozyme hadn't been invented, and hypertonic saline was discussed only in chemistry classes. So, I jogged with the other lemmings, and cleared my lungs out this way.

> *"Beyond the immense psychological benefits, the absolute strength derived from weightlifting has provided countless physical assists in dealing with CF."*

But vanity (I must have had a lot of this) also led me to another form of exercise that was less popular, especially for 20-year-old women. I started lifting weights. And, I got strong...very strong. I first became hooked when I noticed after a few weeks of arm weights that my shirt sleeves were tighter. At first I thought the shirt had shrunk, but then my hand brushed against my upper arm and I noticed it was hard...as in muscular.

JULIE DESCH, MD

Whoa, I thought, this stuff works! Then I started buying books about "women's weight training." I learned all the exercises and started doing them. A few years later, I was bench pressing my weight, and there would be times where I'd look around and realize that I was the strongest woman in the gym. Now, this was a pretty amazing concept to wrap my "I have CF and will never be an athlete" brain around. I still ran, because I started to realize it was good for more than just my pant size, but my fitness passion became all about weight training.

In my experience, there is noth-

ing more empowering than realizing that I can do something physical as well as, or even better than, CFTR-able gym rats. Lifting weights and the resulting strength gains did this for me. The gym became my "CF can't touch this" happy place. Mind you, I am not talking about the "cardio" area of the gym. CF most definitely rules in that place. I'm talking about free weights, squat racks, and yes, even the dreaded barbell deadlift platform. Strutting around the gym doing my thing three or four days a week was and still is a very effective coping mechanism I've fine-tuned after thirty-plus years of lifting while living with a nasty disease.

But beyond the immense psychological benefits, the absolute strength derived from weightlifting has provided countless physical assists in dealing with CF. I think it was the legendary strength coach, Dan John, who developed the metaphor of absolute strength being like a glass (the drinking kind). The bigger the glass, the easier it will be to achieve your fitness goals, whatever they are. He likens absolute strength to a container which holds everything else fitness related (mobility, strength endurance, flexibility, etc.) The bigger the container, the more of these other qualities fit. For example, if I can bench press 100 pounds, there is a very good chance that I can do many more push ups than someone who can press only 50 pounds. I also likely have better shoulder flexibility and mobility.

I like to expand on his metaphor here, and say that the strength container holds many CF care related items as well. For example, if I have developed abs of steel by doing heavy front squats, I am willing to bet my Mobilaire that my cough is stronger and I am able to clear more crap from my lungs than someone who has minimal abdominal strength. If I have increased my thoracic mobility by doing heavy Turkish get ups, or jerks, I know that my chest wall mechanics are optimized for full lung expansion. If I have built up some serious lean mass by lifting for hypertrophy, I know I have improved my glucose metabolism. See what I mean? Strength is a tremendous benefit, not just for your psyche, but also for your ninja CF fighting skills.

Do cardio, by all means. It shakes you up and makes you breathe faster and deeper. This is great for airway clearance. But do NOT neglect weight training and fail to take advantage of the amazing benefits of just being badass strong! ▲

Julie is 53 and ***is a physician who has CF. She may be contacted at: jdesch@usacfa.org***.

TILLMAN *continued from page 13*

S. Jensen-Fangel, H.V. Olesen, M. Skov, L.E. Lemming, T. Pressler, H.K. Johansen, N. Nørskov-Lauritsen. Journal of Cystic Fibrosis. Volume 12, Issue 6 , Pages 638-643, December 2013

In this nationwide retrospective study, the authors analysed species distribution, antimicrobial susceptibility and time to next occurrence of Achromobacter in Danish cystic fibrosis patients from 2000 to 2011. Early treatment with inhaled antibiotics may prevent or postpone chronic infection with Achromobacter in CF patients.

http://tinyurl.com/n4ktvz7

Clinical Use of Tobramycin Inhalation Solution (TOBI) Shows Sustained Improvement in FEV1 in Cystic Fibrosis. Michael W. Konstan MD, Jeffrey S. Wagener MD, David J. Pasta MS, Stefanie J. Millar MS, Wayne J. Morgan MD. Pediatric Pulmonology, Article first published online: 9 SEP 2013

The authors assessed the change in level and trend of FEV1 % predicted over a 2–year period associated with initiation of Tobramycin Inhalation Solution during routine clinical practice. Initiating chronic TIS therapy in the routine clinical care of patients with CF was associated with improvement in FEV1 % predicted but no change in rate of decline, which means that this benefit was sustained over the 2 years studied.

http://tinyurl.com/kucydqu

Enteral Tube Feeding in Adults with Cystic Fibrosis; Patient Choice and Impact on Long Term Outcomes. H. White, A.M. Morton, S.P. Conway, D.G. Peckham. Journal of Cystic Fibrosis. Volume 12, Issue 6, Pages 616-622, December 2013

Supplemental enteral tube feeding (ETF) improves clinical outcomes when administered over 3years, resulting in significant weight gain, a normal BMI and stabilization of lung function. It does not reduce intravenous antibiotic treatment days. In contrast those patients eligible for, but who declined ETF, showed a deterioration in lung function and a failure to gain weight and to achieve normal BMI status.

http://tinyurl.com/n37559g

Poly-l-lysine Compacts DNA, Kills

Continued on page 23

COUGHING WITH A SMILE...

Through my Eyes

By Jennifer Hale

Hello, *CF Roundtable* readers! It has been way too long. I have been going through my own personal challenges the last five months, but I am back to writing my column. While I have been down for the count I have watched a lot of movies. I have been on IVs for the last six months and have most likely seven or more months to go.

I know you are probably thinking, whaaaat? I know I am thinking that and I am living it. I have never been on IVs longer than three weeks in my 41 years, and that was only one or two times. So, to think I have doing it for the last six months is mind boggling, but I am getting through it.

I am fighting a bacterium called Mycobacterium abscessus (M. abscessus) and let me tell you this bacterium is relentless! My doctor tells me it can be eradicated, so I am hoping I am one of the lucky ones who shows this bacterium the door!

As I stated earlier, I have been watching a lot of movies and one in particular called "The Curious Case of Benjamin Button", starring Brad Pitt, has struck a chord with me. What struck that chord, in particular, is a quote that Brad Pitt's character, Benjamin, says to another character in the story, whose name is escaping me. (I am supposed to be writing about "Memory Problems" for this issue so there you go - there is an example of my forgetfulness. LOL!) Anyway, Brad Pitt is asked what it is like growing younger and his response is, "I don't know, I'm always looking out my own eyes."

I thought that was profound in the aspect of relating to CF too. I do not know what it is like to be healthy, to breathe freely, to not struggle day in and day out and for me that is my norm. I am always breathing through my own lungs, doing my therapies and most importantly adjusting my sails to all the ups and downs of this disease. I adjust so much and all the time that I think I do not even realize how hard my life is or how unhealthy I might be. I just do and I just am. I am just looking through my own eyes and to me this is my norm-to fight, to adjust and to keep moving forward.

Then I thought to myself, what makes some people able to gather up strength and keep persevering even though they are getting kicked every step of the way and others are crippled by their circumstance? As I think about this, I am not really sure why some people can handle adversity and others just crumble. I think it is a conscious choice to keep putting one foot in front of the other. I also feel one needs positive people around them, cheering them on and helping them along the way. As I stated in previous columns my husband is my positive ray of sunshine, for sure. I also think it has a lot to do with what one of my favorite preachers, Joyce Meyer, has said, "Circumstances can't dictate the way you live; you must be stable in all circumstances good and bad." I think that is a great point and what I try to do in my life. The CF journey has lots of bad times but that does not mean I have to be a sour, pessimistic and angry person. I feel being that way will not change the circumstances so what is the point?

JENNIFER HALE

The last three years my strength and perseverance have been tested. I have gone from an FEV_1 of 65% to as low as 31%, lost 17 pounds, had a port placed in my chest , IV infusions daily and have to use O_2 when exercising. It has been an absolute roller coaster. The last six months were my lowest in FEV_1 and weight and I feared that this was the beginning of the end. I actually, for the first time in my life, was scared CF was killing me and the road to transplant or death was coming over the horizon.

Did you know the definition of horizon is just the apparent intersection of the earth and sky as seen by the observer? Therefore, the horizon is subjective and can keep moving based on the observer. With that said, during the bleakest of times the horizon is up to you. It can be close and be a signal for positive change or you can see it as being far, far away and there is still time for everything to turn around. Never give up!

My M. abscessus journey has led me to getting a port. I really had no

choice in the matter because I was going to have to do IVs for 12 to 18 months and it would be easier with a port then a PICC line. So I got my port and because I am so skinny you really can see it in my chest and the catheter going up my neck. But it has worked out really well and it is super easy to do IVs and my arms are free. Getting a port was emotional for me because it was again the road to thinking I was declining rapidly. I was becoming the skinny, port in chest, 0_2 toting CFer and I just never viewed myself that way nor thought that I would ever get to that point. That was not me! I am not that kind of CF person.

Yet again, I had to adjust my sails. But let me tell you what a number it does on your psyche having a port with tubing hanging from your chest and O_2 tubes in your nose while you are trying to work out at the gym. It has been hard to go to gym in that state, but I need the gym and I will never stop going. I sometimes think with all my paraphernalia hanging off me and my huffing, puffing and coughing someone might be thinking, please girl just go home and rest! But I hope they are thinking what a strong woman and really I am trying to not even care what they are thinking.

I once read that you would be surprised how much people around you are not thinking or paying any attention to you at all. I have really learned to not care what people think and to not let others keep me from being healthy and moving forward. If you think about it, why let someone keep you from doing what is right and what is fun and what is good for your health? Who cares what others think? It's all about what you need to do to get healthier! So strap those shoes on and get out there. Know that small steps are better than no steps at all.

I leave you with this great quote that Jerry Cahill posted on Facebook by an unknown:

I can choose to let it
Define me
Confine me
Refine me
Outshine me
Or I can choose to
Move on and leave it
Behind me

Till next time, *CF Roundtable* readers, I gotta go to the gym before my next IV infusion! ▲

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

TILLMAN *continued from page 21*

Bacteria, and Improves Protease Inhibition in Cystic Fibrosis Sputum. Alice V. Dubois, Patrick Midoux, Delphine Gras, Mustapha Si-Tahar, Déborah Bréa, Sylvie Attucci, Mustapha-Kamel Khelloufi, Reuben Ramphal, Patrice Diot, Francis Gauthier, and Virginie Hervé. American Journal of Respiratory and Critical Care Medicine. Volume 188, Issue 6

The authors examined whether poly–l–lysine would compact DNA in cystic fibrosis lung secretions and liquefy CF sputum, improve the control of extracellular proteases by exogenous inhibitors, and whether it displays antibacterial properties toward CF–associated bacteria. Poly–l–lysine may be an alternative to dornase–αto liquefy sputum with added benefits because it helps natural inhibitors to better control the deleterious effects of extracellularly released neutrophil serine proteases and has the ability to kill bacteria in CF sputum.
http://tinyurl.com/qfsjva3

Ivacaftor treatment of cystic fibrosis patients with the G551D mutation: a review of the evidence. Kavitha Kotha, John Clancy. Therapeutic Advances in Respiratory Disease. Published online before print September 3, 2013

Ivacaftor treatment produces dramatic improvements in lung function, weight, lung disease stability, patient-reported outcomes, and CFTR biomarkers in patients with CF harboring the G551D CFTR mutation compared with placebo controls and patients with two copies of the common F508del CFTR mutation. The unprecedented success of ivacaftor treatment for the G551D CF patient population has generated excitement in the CF care community regarding the expansion of its use to other CF patient populations with primary or secondary gating defects.
http://tinyurl.com/k99lnv7

Inhalation Solutions — Which Ones May Be Mixed? Physico-chemical Compatibility of Drug Solutions in Nebulizers. Wolfgang Kamin, Frank Erdnüss, Irene Krämer. Journal of Cystic Fibrosis. published online 30 October 2013.

Many patients suffering from chronic respiratory diseases rely on inhalation therapy with nebulizers. About 25% of patients who need to inhale several different drugs per day

Continued on page 27

IN THE SPOTLIGHT

With Nathan Wiebe

By Jeanie Hanley with Andrea Eisenman

Nathan Wiebe (pronounced "we be") is passionate about life and good-natured. He has lived for 34 years, not always in good health. After difficult teenage years, he decided to take control of his life and his health. His experiences have motivated him to become the successful business man he is today, pursuing a career in investigating fraud in business practices, going after corrupt politicians and city officials. He has been married for almost 10 years to Molly, whose life is quite an inspiration too. His devotion to swimming has served as the cornerstone for excellent health. After this interview it was easy to understand why he is fully committed to all of his endeavors. Nathan shared a great deal of his inspiring life history with humor and sheer honesty. Please welcome our latest star. Spotlight please!

How were you diagnosed with CF?

I was diagnosed at three months of age. My parents and I were living in Lafayette, Louisiana at the time. Due to the warmth in the weather I apparently had a difficult time retaining electrolytes, and ultimately stopped eating and became lethargic.

My parents then had me hospitalized in a local hospital where the doctors could not determine my diagnosis. I was ultimately transported to the University of Texas Hospital at Galveston where I was diagnosed using a sweat test.

What were the main issues as a child?

My primary issues as a child were gastrointestinal in nature, as well as electrolyte. I was not really symptomatic with my lungs until I became a teenager. As a teen I developed the usual sputum cultures and my CF battle advanced to my lungs.

NATHAN WIEBE IN VANSBRO, SWEDEN, 2012, IN A RACE WHERE HE PLACED 143RD IN THE MENS EVENT OUT OF APPROXIMATELY 5000.

What was life like growing up with a sister who also had CF?

My younger sister, Meara, on the other hand, seemed to have more lung issues during her childhood. I recall her using a nebulizer regularly as a child. We used to go to clinic visits at the Cystic Fibrosis Center at Children's Hospital Los Angeles together. It was tough seeing my sister sick, but she has the spirit of a fighter.

What are your symptoms now?

I have chronic infection in my lungs, mucus production, digestive issues, high blood pressure, cystic fibrosis related diabetes, and fairly regular hemoptysis.

I have a routine that I follow religiously which is exercise (usually swim), hydrate, nebulize hypertonic saline, clearance in the shower with the Quake (an airway clearance device), nebulize pulmozyme, then the Tobi Podhaler (if I am on cycle). Oh, and the enzymes, multivitamins, blood pressure meds, azithromycin... I am sure that I am forgetting something here.

How is your sister now?

My sister is post-op double lung transplant, and is doing well, all things considered. She lives in San Diego with her veterinary doctor husband, Jeff, and their numerous animals.

What are your CF genetic mutations?

DeltaF508 mutation (homozygous)

What do you do for a living?

I am the Supervising Senior Investigator for the City of Austin City Auditor. I investigate fraud, waste and abuse across the city. We have an investigative team composed of highly competent, wonderful people who have a strong desire to serve the public. We have investigative purview over City operations which include approximately $3 billion in expenditures annually, 40 Departments, and 15,000 employees.

How did you decide on this career?

I have an undergraduate degree in Information Systems from California State University, Los Angeles. I began my career as an information systems consultant and worked for a "white-collar" forensics firm. I ultimately obtained an MBA from the University of Southern California and became a partner of the firm.

When my wife applied for and received admission to a PhD program in Education at the University of Texas at Austin, we relocated to Texas, where I began work for the

Federal Deposit Insurance Corporation as an investigator of bank fraud by financial institution insiders.

Ultimately the travel requirements of the job began to conflict with my health and I found my current position with the City of Austin.

Were you always so motivated, even as a teenager?

I was highly motivated to slack off as a teen. Does that count?

What happened during your teenage years?

I believe it was half denial and half rebellion. Being a teenager in school where you perceive that everyone is watching what you're doing, and then having to go to the nurses office at lunch to take your medications isn't a formula that adds up to "ideal" for a teen. Additionally if I was out with friends and didn't have a car, having to derail the events by going home to get my medications was not an option for me at that point. Bottom line, upon reflection, I was an idiot.

What or who turned you around as a teenager?

Well there were a number of things, the first being in my late teens I attended an international school in Denver Colorado. There were individuals from around the globe who came to the US to learn English as well as the U.S. culture. I became friends with a number of individuals who have overcome significant challenges in their backgrounds. These individuals inspired me, and though they didn't have cystic fibrosis, their challenges weren't any less than my own.

Secondly, I had always had strong support from my family. My parents and my sister all were experienced in dealing with cystic fibrosis and all of the various elements that come along with that.

Lastly, and most importantly, I met the love of my life, Molly. Molly is a first-generation refugee who was born in Cambodia during the time of Khmer Rouge genocide. She came to the US as a young child with her parents with almost no possessions. Her parents had the equivalent of elementary education, and spoke no English. Despite all of that Molly was a strong individual who had a passion for life and very little patience for allowing excuses to stand in the way of achieving her goals. She rocked my world.

I drew strength from all of these things, and thankfully, was able to make a change and begin challenging life head-on rather than reacting to it, as tumbleweed would react to the winds.

How did you build up exercise tolerance after having very little?

Shortly after meeting Molly, I returned to the sport of my childhood, swimming. Given the fact that I hadn't taken care of myself for a number of years, this was no easy task. I began by spending more minutes in the pool. Slowly over the course of years, I added distance and time to my workouts.

Also around this time I became interested in triathlons. Having success at building up some capacity at swimming, I approached running and biking with the same methodology. I ultimately completed the Los Angeles Triathlon in two different years. Since that time I have focused more on swimming as a sport and less on running and biking.

Where do you swim?

I'm currently a member of the Texas Longhorn Aquatic Masters (TXLA) swim club. There I am coached by a world-class coach, Whitney Hedgepeth, who is the 2013 United States Masters Swimming Coach of the Year. I usually make four practices a week, where we swim around 3+ miles per workout. I am in the best shape of my life and my PFTs show it.

What impact has CF had on your career? Relationships?

This is a tough question, since there are ways that CF has factored into my life in unconscious ways in addition to the ones that I am aware of. That said, CF has had a great impact on my life to my knowledge.

Besides the obvious things like always needing adequate health insurance, life insurance, and adequate time off in case I get sick, I find that CF gives me the knowledge that my time on earth may be limited and I need a profession that will leave a positive mark on society. Thus, in choosing a career I found myself drawn to serving the public.

By the same token, CF has impacted my relationship with my wife. It has filled me with the desire to make her happy every day in every way that I can.

How did you meet Molly?

I met Molly one day when I was at a friend's house playing pool. Molly's younger sister was friends with one of my friend's younger sisters, so Molly just happened to be picking her up that day from my friend's house while I was there. What a stroke of luck. When retold by my sister-in-law and friends, the story goes that when Molly walked in the room I stood up and physically moved people out of the way to get to Molly to introduce myself. That's not the way I recall it; however it's my word versus a number of other people's, so I guess I'm out of luck.

How do you fit it all in -exercise, work and marriage? What's your day-to-day routine?

Well I think I am in no way unique in having a whole lot to do. My typical day begins with me getting up at 5:30, drinking a breakfast shake and some coffee, and heading to the pool to workout for an hour and a half. I then head to the office where I spend my days fighting fraud, waste and abuse with a team of fantastic people. I usually come home at around

Continued on page 26

five and do my nebulizer, airway clearance with the Quake, and Pulmozyme routine. I then cook dinner for Molly and me. I usually have some free time after this, but currently I'm taking a class and that eats into my evening for about two-and-a-half hours two times a week.

Yeah, sure – that's what I do everyday too. Anyway, have you ever considered having children?

I have considered having children and that is something that Molly and I are currently debating. On one side of the coin, with the hand I've been dealt, Molly could be raising children alone. On the other, who knows what the future will bring. Such is life.

Are you religious?

No I'm not religious. However I do practice mindful meditation, and have worn a jade Buddha since my mother-in-law gave it to me almost 15 years ago, and believe in doing to others as you would have them do to you.

How did your sister having a double-lung transplant affect you?

My sister's double lung transplant was incredibly stressful for the entire family. Actually, the year leading up to the transplant was the most difficult. Meara almost didn't make it. At UCLA she was placed on ECMO a number of times, and we were lucky that she made it through.

We in the cystic fibrosis community get the pleasure of meeting and knowing some wonderful people, so when seeing someone with CF facing these obstacles, we have the choice to stand tall and try to lend our strength, or despair. Or maybe we just do our best to walk a path between these two.

After your father passed away, what helped you deal with his loss?

Family, friends and coworkers. Regular exercise and good food didn't hurt either.

For you, what is the hardest part of having CF?

Not letting the fact that we don't have control over certain things overshadow how much control we do have in our lives.

Any benefits of having CF?

Absolutely, seeing every day is a miracle. Every day that I wake up and see my wife, go to swim practice, spend time with family and friends, and live life is colored with these lenses that are cystic fibrosis.

Ever participated in a CF Clinical Trial and, if yes, would you recommend it?

Yes, I have participated in a number of CF clinical trials, and I strongly advocate anyone who has the opportunity to do so. It is fantastic that we have a number of drugs in the pipeline that have the potential to change the game for cystic fibrosis. Volunteering for clinical trials ensures that these will continue to come to market and that the community will ultimately receive the benefits of all the hard work that has gone into them.

Funniest CF moment?

The moment that I realized that many of the medical practitioners at the CF clinics are younger than I am.

Who do you admire the most?

My wife, absolutely. She's currently working on her PhD in education at the University of Texas. This is a woman who is the first of her family to go to college. Plus, she puts up with me!

Additionally there's my mom. Through my father passing away from cancer and my sister and myself having CF, and all the other nuances of life, she has remained a strong and positive person. That strength and positivity is something that I try to hold in my own life.

What goals are you currently working toward?

Here are my major to-do items, not necessarily in order: 1) Provide as many smiles to my loved ones as possible. 2) Further enhance my ability to support my family and friends. 3) Continue to advance in my career, and protect and serve the public. 4) Get faster in the pool!

What are qualities that you value most in a person?

Love of life, seizing the day, and showing respect and kindness to those around you.

Is there anything you wish you could do over again?

No. Though I have certainly made some foolish decisions, as all people have, there is no place that I'd rather be than here at this place and moment in my life. ▲

Jeanie Hanley is 51 and is a physician who has CF. She is a Director of USACFA and is the Vice 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.

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save time by mixing them for simultaneous inhalation. This review presents a comprehensive overview of the available data concerning physico-chemical compatibility of commonly mixed nebulizer solutions and suspensions. Results indicate that many nebulizer solutions/suspensions are mixable without provoking incompatibilities. However, certain inactive ingredients contained in some of the tested drug products could be identified as a reason for incompatibilities, e.g. impaired activity of dornase alfa. The clinical efficacy of simultaneous inhalation of duplicate, tripartite or quadripartite mixtures must be evaluated in clinical studies before final recommendations for the inhalation regimens can be made.
http://tinyurl.com/m6cavpa

BACTERIA/FUNGI/VIRUSES

Factors Influencing the Acquisition of *Stenotrophomonas maltophilia* Infection in Cystic Fibrosis Patients. Sanja Stanojevic, Felix Ratjen, Derek Stephens, Annie Lu, Yvonne Yau, Elizabeth Tullis, Valerie Waters. Journal of Cystic Fibrosis. Volume 12, Issue 6 , Pages 575-583, December 2013

This study illustrates the evolution of S. *maltophilia* infection over time in a large cohort of adults and children with CF. Younger CF patients, and those with greater lung function decline were at increased risk of S. *maltophilia* infection. The use of oral antibiotics to maintain lung function may be a way of decreasing the risk of infection. However, the optimal management of CF patients with persistent S. *maltophilia* infection is not yet known and requires further studies.
http://tinyurl.com/pp2q98l

Achromobacter species in Cystic Fibrosis: Cross-infection Caused by Indirect Patient-to-Patient Contact. C.R. Hansen, T. Pressler, W. Ridderberg, H.K. Johansen, M. Skov. Journal of Cystic Fibrosis. Volume 12, Issue 6 , Pages 609-615, December 2013

Achromobacter species can cause cross-infection even after a short period of indirect contact between infected and non-infected CF patients. Patients should be followed closely for several months before the possibility of cross-infection is ruled out.
http://tinyurl.com/mvl7d5h

The Basidiomycetous Yeast *Trichosporon* May Cause Severe Lung Exacerbation in Cystic Fibrosis Patients – Clinical Analysis of *Trichosporon* Positive Patients in a Munich Cohort.

Continued on page 28

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 & fuller lives, and by generous support from Genentech.

TILLMAN *continued from page 27*

Carolin Kröner, Matthias Kappler, Ann-Christin Grimmelt, Gudrun Laniado, Benjamin Würstl and Matthias Griese. BMC Pulmonary Medicine 2013, 13:61

This study demonstrates the potential association of *Trichosporon* spp. with severe exacerbations in CF patients.
http://tinyurl.com/meoxqfv

Nontuberculous Mycobacteria: The Changing Epidemiology and Treatment Challenges in Cystic Fibrosis. Leung, Janice M., Olivier, Kenneth N. Current Opinion in Pulmonary Medicine: November 2013 - Volume 19 - Issue 6 - p 662-669

Given increasing prevalence rates, clinicians should maintain a high level of suspicion for NTM as disease-causing organisms in CF, particularly for M. *abscessus*. New knowledge regarding this species, however, can help to tailor appropriate therapy.
http://tinyurl.com/msrbfjq

Incidence and Clinical Impact of Respiratory Viruses in Adults with Cystic Fibrosis. William G Flight, Rowland J Bright-Thomas, Peter Tilston, Kenneth J Mutton, Malcolm Guiver, Julie Morris, A Kevin Webb, Andrew M Jones. *Thorax*. Published Online First 14 October 2013

Viral respiratory infection is common in adults with CF and is associated with substantial morbidity. Respiratory viruses are a potential therapeutic target in CF lung disease.
http://tinyurl.com/lqbxa88

Epidemiology of Nontuberculous Mycobacterial Infections and Associated Chronic Macrolide Use among Persons with Cystic Fibrosis. Binder A, Adjemian J, Olivier K, Prevots D; American Journal of Respiratory and Critical Care Medicine (Aug 2013)

Persons with CF are at high risk of nontuberculous mycobacterial (NTM) infection, with treatment requiring prolonged multi-drug regimens that include macrolides. While macrolides, specifically azithromycin, are used in the management of CF patients with chronic Pseudomonas, macrolide-resistant NTM infections are of growing concern. Patients with incident NTM infections from either MAC or M. *abscessus* were less likely to have had chronic azithromycin treatment in the past year. However, because macrolide mono therapy may lead to macrolide resistance, routine screening for NTM should be considered for persons with CF.
http://tinyurl.com/laygpcf

Inflammatory and Immunological Biomarkers are Not Related to Survival in Adults with Cystic Fibrosis. K.L. Moffitt, S.L. Martin, A.M. Jones, A.K. Webb, C. Cardwell, M.M. Tunney, J.S. Elborn. Journal of Cystic Fibrosis. published online 15 July 2013.

Chronic *Pseudomonas aeruginosa* pulmonary infection is associated with a decline in lung function and reduced survival in people with CF. Damaging inflammatory and immunological mediators released in the lungs can be used as markers of chronic infection, inflammation and lung tissue damage. The data indicate that biomarkers of inflammation are not independent predictors of survival in people with CF.
http://tinyurl.com/m8rb3q3

FYI

Diabetic Myonecrosis in a Cystic Fibrosis Patient. Benjamin T Kopp, MD, Stephen Kirkby, MD, Don Hayes Jr, MD, Kevin M Flanigan, MD. Respiratory Care October 1, 2013 vol. 58 no. 10

Cystic fibrosis related diabetes is an increasingly common comorbidity in CF patients, with scarce data on end–stage complications in the CF population. The authors report the case of a 32–year–old with poorly con-

trolled diabetes presenting with sub–acute leg pain and focal quadriceps tenderness. Neuromuscular testing and extensive workup revealed diabetic myonecrosis. This is the first reported case of diabetic myonecrosis in a patient with CF, and highlights the need for pulmonary physicians to recognize this diabetic complication in CF patients, which is associated with a poor long–term prognosis and existing microvascular complications.
http://tinyurl.com/o437527

Predictors for Future Cystic Fibrosis-related Diabetes by Oral Glucose Tolerance Test. Kerstin Schmid, Katharina Fink, Reinhard W. Holl, Helge Hebestreit, Manfred Ballmann. Journal of Cystic Fibrosis. Published online 27 June 2013.

An annual oral glucose tolerance test (OGTT) has become part of standard care in CF to screen for CF-related diabetes (CFRD). The objective of this study was to determine predictors for future CFRD derived from an OGTT. In this large study, impaired fasting glucose, impaired glucose tolerance, and indeterminate glucose tolerance were all predictors of future CFRD. The OGTT in patients with CF should include a 1-hour post-challenge value.
http://tinyurl.com/kb6wuvz

Cystic fibrosis and Pregnancy in the Modern Era: A Case Control Study. Monica Ahluwalia, Jeffrey B. Hoag, Anas Hadeh, Marianne Ferrin, Denis Hadjiliadis. Journal of Cystic Fibrosis. published online 09 September 2013.

Increasingly, women with CF become pregnant. Outcomes of these women need further study, particularly in the setting of improved survival in CF. Pregnancy does not lead to immediate or medium-term adverse effects for CF patients.
http://tinyurl.com/on5qrjo

Chest Physiotherapy Compared to No Chest Physiotherapy for Cystic Fibrosis. Louise Warnock, Alison Gates, Cees P van der Schans. Editorial Group: Cochrane Cystic Fibrosis and Genetic Disorders Group. Published Online: 4 SEP 2013

The lungs of people with cystic fibrosis produce excess mucus. This leads to repeated infection and tissue damage in the lungs. It is important to clear the mucus using drugs and chest physiotherapy. Physiotherapy clears mucus by different techniques or by using mechanical devices or both. Daily physiotherapy takes a lot of time and trouble. The authors summarize the findings of eight studies; they found that methods of clearing the airways have short-term benefits for moving mucus. Three studies measured sputum which had been coughed up and found a higher amount with chest physiotherapy; four studies measured radioactive tracer clearance and found increased clearance with chest physiotherapy. Only one study reported an improvement in lung function in some of the treatment groups; but three other studies who reported this outcome did not find any significant effect from chest physiotherapy. At present there is no clear evidence of long-term effects in chest clearance, quality of life, or survival with chest physiotherapy.
http://tinyurl.com/kxolggj

Sinonasal Manifestations of Cystic Fibrosis: A Correlation Between Genotype and Phenotype? M.C. Berkhout, C.J. van Rooden, E. Rijntjes, W.J. Fokkens, L.H. el Bouazzaoui,

Continued on page 30

H.G.M. Heijerman. Journal of Cystic Fibrosis. Published online 07 November 2013.

Patients with CF are prone to develop sinonasal disease. The prevalence of rhinosinusitis in adult patients with CF was 63% and the prevalence of nasal polyps 25%. Patients with class I–III mutations had significantly smaller frontal sinuses, sphenoid sinuses, more opacification in the sinonasal area and more often osteitis/neoosteogenesis of the maxillary sinus wall compared to patients with class IV and V mutations.
http://tinyurl.com/kgj4w7z

Cystic Fibrosis-related Bone Disease: Insights into a Growing Problem. Stalvey, Michael S.; Clines, Gregory A. Current Opinion in Endocrinology, Diabetes & Obesity. December 2013 - Volume 20 - Issue 6 - p 547-552

Cystic Fibrosis-related bone disease (CFBD) continues to increase as the life expectancy of individuals with CF increases. According to clinical guidelines, individuals with cystic fibrosis should be initially screened at the age of 18 years via dual-energy x-ray absorptiometry, if not done previously. The underlying pathogenesis of CFBD appears to be multifactorial, but increasing data imply a direct impact by the cystic fibrosis transmembrane conductance regulator (CFTR). CFTR deficiency and/or dysfunction impair osteoblast activity and differentiation, and indirectly promote osteoclast formation. Unfortunately, once diagnosed with CFBD, few cystic fibrosis tested medical therapies exist.
http://tinyurl.com/malpze6

Xbox Kinect Represents High Intensity Exercise for Adults with Cystic Fibrosis. Hayley Holmes, Jamie Wood, Sue Jenkins, Peta Winship, Dianne Lunt, Susan Bostock, Kylie Hill. Journal of Cystic Fibrosis. Volume 12, Issue 6, Pages 604-608, December 2013

Exercise is important for patients with CF. Interactive gaming consoles are a new trend in exercise. This study sought to determine the exercise intensity of training using the Xbox Kinect. Training using the Xbox Kinect represents high intensity exercise for adults with CF and may be a suitable alternative to conventional exercise modalities.
http://tinyurl.com/otyza7b ▲

Laura Tillman is 66 and has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.

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Genetic Mutation Information Resource

Collaborators from several institutions around the world and the US Cystic Fibrosis Foundation are excited to announce that a new resource – CFTR2 – is now available for public use! This is the result of an international research collaboration to provide information about specific cystic fibrosis gene mutations to patients, their families, researchers, health professionals, and members of the general public. We hope that you will find the information useful.

The website is available at www.cftr2.org. Once you have reviewed the website, please take a few minutes to complete the user satisfaction survey located in the blue box "How can you help us improve the website?" in the "Quick Links" section of the left margin. Your responses will help us improve the website. Please feel free to contact cftr2@jhmi.edu with any comments, questions, or suggestions; but, please note that we are unable to answer any questions about the medical care of individual patients, since we are the research team that helped develop the website and not a clinical care team.

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her relationships with the staff at Johns Hopkins Hospital – where it seemed she spent half her life. She loved the doctors and nurses who worked with her. In turn they loved her back and respected her almost incredible knowledge of her disease and her own body. She displayed that same affection and respect for all the other staff such as aides, cleaners, meal servers, etc. Kathleen learned about many of their families, sincerely asking about them often. She unfailingly thanked every person for what they did for her.

I was privileged to be married to Kathleen. She taught me so much by the example of her life which she lived so well. Mostly though, she loved me and, like everyone else, I loved her back. ▲

Sincerely,
Dick Harris
Bowie, MD

REMINDERS

- Please notify us immediately of any address changes. Returned mail wastes money and delays mailings.
- We would like to act as a referral source for active adult support groups. Please send us your group name, leader's name and phone number, number and age range of your members and geographical area covered, and we will add you to our referral list.
- Please let us know of the major occurrences in your life (*e.g.*, *marriages*, *births*, *completion of educational degrees or training*, *career advancement*, *transplants*, *anniversaries*, *birthdays*) and we will print your information in **Milestones**.
- Share your ideas for **Focus Topics**, feature articles or any suggestions for improvements you may have to help make *CF Roundtable* more relevant and interesting to you.
- You can reach **USACFA** and ***CF Roundtable*** at anytime by phone **(248) 349-4553**. You may email us at **cfroundtable@usacfa.org**
- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq., 712 Main, Suite 2130, Houston, Texas 77005. Email: cflegal@cff.org**.
- You may subscribe at www.cfroundtable.com

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IMPORTANT RESOURCES

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

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

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

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

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