CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS SUMMER 2011

Tonic and Me: Traversing the Winter From Hell

By Julie Desch

There is good news and bad news about getting old with cystic fibrosis. The good news is that I am getting old with CF! How great is that? Although it is happening to more and more people as medical care gets better and the median age of survival creeps upward, it still surprises me every time I have a birthday. I laugh at my wrinkles, chuckle at hot flashes, and marvel at the fact that I look, well, middle aged.

Living with an unpredictable illness is not all mirth and laughter, though. Some aspects of getting older are a bit more frustrating, at least for me. This is where "Tonic", a new iPhone/iPad app, enters my story. As you probably know, staying healthy with CF requires more and more effort with age. There are more medications to take, more treatments to do, more effort is required to stay fit, to eat well, to manage CFRD, to get enough sleep, enough water, to do whatever it takes to make the plumbing system work well, remember appointments for the doctors and port flushes, to do...pretty much everything. Yet, just as the "CF care complication" factor increases, the ability of the mind (mine, at least) to keep track of it all begins its downward descent. This is poor design, but it is what it is.

I noticed it first with Advair. I can never remember if I have taken it. I don't know... I seem to have a mental block. The block then spreads to enzymes, vitamins, children's names, books I've read, etc. While it's much cheaper to be able to read the same book (and be entertained) multiple times, too many shots of ProAir or too many Pancrelipase capsules can be problematic.

I have officially dubbed this past winter, "the winter from hell". For some reason, I had three episodes of pneumonia with resulting rounds of home IV antibiotics over the span of eight months. If this wasn't enough, I cultured MRSA recently, so each round of IVs included three different medications, all through IV access. IV meds must be refrigerated, of course, and when they come in Entermates (as two of mine did), they need to be removed from the refrigerator a few hours before being used. When you forget to do this, 1) brrrr, and 2), the infusion takes forever. Not to mention that during exacerbations, I do three treatments per day, try to sleep, try to remember to eat (when I have no appetite), and take the usual oral meds, supplements, and vitamins. I also have follow up appointments with doctors

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INSIDE THIS ISSUE

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

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EDITOR'S NOTES

Welcome to our summer issue of *CF Roundtable*. One problem for me during these hot months is staying hydrated enough to enjoy outdoor activities. Drinking coconut water helps me. It keeps my electrolytes stable and I don't experience hyponatremia or dehydration. See the blog on our website: http://www.cfroundtable.com/2011/06/10/cuckoo-for-coconut-water/

Our Focus topic is: "If Only I Had Known Then What I Know Now". It garnered many articles. See what **Debra Radler, Laura Mentch, Jen Eisenmann, Nicole Matthews,** and **Jeanie Hanley** have to say about learning from the past and knowing better now. See **Kathy Russell's** "Speeding Past 50" for what she knows now but did not have available to her many years ago. In "Wellness", **Julie Desch** writes about what she would tell herself at various stages of her life. Similarly, **Debbie Ajini** uses "A Deep Breath In", to advise herself at different ages in her life to get through the tough times, noting better times ahead. In "Transplant Talk", **Colleen Adamson** and **Andrea Eisenman** share their experiences with skin cancer while being on immuno-suppressants.

As always, **Laura Tillman** compiles news that she finds online about the newest treatments and interesting factoids for and about CF in "Information from the Internet". **Beth Sufian** answers questions about high co-pays, COBRA, SSI and SSDI in "Ask the Attorney". In "Spirit Medicine", **Isa Stenzel Byrne**s examines the difficulty one with CF might feel in receiving from others and writes that it is part of a cycle of give and take. In "Voices from the Roundtable", **Cynthia Dunafon** writes about her trip to The Grand Canyon and **Bonnie Bleiweiss** writes on being newly diagnosed at 58. Bonnie also shares a cartoon on how hectic packing for vacation can be. In "Conversation Corner", **Walter Bartholomew** shares his thought on fighting CF daily. If you enjoyed the first edition of "Taking Flight, Inspirational Stories of Lung Transplant", compiled by **Joanne Schum,** be sure to read the review of the second edition which is now available.

We have an article from **Kathleen East** who started The Blooming Rose Foundation in order to help others through the maze of having CF, whether you are a newly diagnosed patient or a seasoned pro looking for answers (from physicians, caregivers and other adults with CF). Laura Tillman writes about new changes to USACFA and welcomes **Mark Levine,** our newest Director. You can read about him on page 15.

We want to share some exciting news about "The Power of Two", the Movie. It will be screening starting in August, see page 5 for more details. There is a new online nomination form for "Heroes of Hope™ Living with CF". Now it is easier than ever to nominate someone. We are happy to announce a new column called, "In the Spotlight". See page 18 for more information.

Keep cool and stay happy and healthy!

Andrea

Publication of CF Roundtable is made possible by donations from our readers and grants from Sustaining Partners - CF Services and a bequest from the estate of Pamela P. Post in honor of Kathy Russell.

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

Laura Mentch
Bozeman, MT
58 on May 21, 2011

Debra Radler
Roselle, IL
49 on May 31, 2011

Wedding

Colleen & Scott Adamson
Alexandria, VA
14 years on June 28, 2011

Debbie & Louie Ajini
Shelby Township, MI
16 years on June 17, 2011

Jeanie & John Hanley
Manhattan Beach, CA
25 years on June 28, 2011

Arthur & Brandie Herron
Sacramento, CA
3 years on June 21, 2011

Debra Radler & Adrian Gulinski
Roselle, IL
4 years on June 8, 2011

Transplant

Colleen Adamson, 42
Alexandria, VA
Bilateral lungs
13 years on July 3, 2011

Anthony Panzara, 43
Chesapeake, VA
Bilateral lungs
1 year on July 3, 2011

NEW BEGINNINGS

Kellisa C. Myers, 23
Lakewood, CA
Received a BA in Sociology and a BA in Women's, Gender, Sexuality Studies from California State University Long Beach
May 26, 2011

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 topics you might like writing about. In addition, humorous stories, articles on basic life experiences, short stories, art work, cartoons, and poetry would be greatly appreciated. We require that all submissions be original and unpublished. With your submission, please include a photo of yourself (as recent as possible) as well as your name, address and telephone number. Photos will be returned. Send all submissions to: **CF Roundtable, PO Box 1618, Gresham, OR 97030-0519.** Or E-mail to: **cfroundtable@usacfa.org**

Summer (current) 2011: If Only I Had Known Then What I Know Now.

Autumn (November) 2011: CF, It's Not Just For Children. (Submissions due September 15, 2011.) The face of CF has changed over the years. Now many people live into their 30s, 40s, 50s and beyond. Tell us about dealing with the problems of aging when you have what many consider to be a "pediatric" disease. Tell us how CF is different for you, now that you are an adult.

Winter (February) 2012: Our Pets And How They Affect Our Lives. (Submissions due December 15, 2011.) Do you have a pet or pets? How has having a pet affected your life? Does a pet affect CF? What are the positive and negative aspects of having a pet?

Spring (May) 2012: Are You An Optimist or A Pessimist? (Submissions due February 15, 2012.)

ASK THE ATTORNEY

Questions and Answers

By Beth Sufian, Esq.

We continue to receive calls and e-mails about Social Security Administration ("SSA") benefits. This column will answer those questions and a few others.

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 families and their CF Care Center teams. The Hotline is proudly sponsored through a grant from the CF Foundation. The Hotline can also be reached by e-mail at CFlegal@cff.org.

1. How can I obtain assistance with high co-pays for medications used to treat CF?

Many drug companies have different programs to assist patients with issues related to high co-pays. The CF Patient Assistance Foundation (CF PAF), a subsidiary of the CF Foundation, is an excellent resource for information about co-pay assistance programs. The CF PAF can also provide co-pay assistance for certain medications. The CF PAF can be reached at 1-888-315-4154 or on their Website www.cfpaf.org

BETH SUFIAN

2. How can I get 29 months of COBRA if I leave work and become eligible for Social Security benefits?

If a person stops work and applies for Social Security Disability Insurance (SSDI), she should notify her employer that she has left work to apply for Social Security Administration (SSA) benefits. A person will want to COBRA her insurance benefits if she has no other options for coverage, such as becoming insured under a spouse's policy. The person needs to COBRA the benefits because it will take 29 months, after the first full month the person is disabled, for the person to become eligible for Medicare. Often people are told the Medicare waiting period is 24 months long. However, if you add the first 5-month waiting period before SSDI benefits begin, it is really 29 months. A person who leaves work for any reason typically is eligible to COBRA his/her insurance benefits for 18 months. If the person meets certain requirements the person will be eligible for an additional 11 months of coverage under COBRA. At the end of the 11 month extension the person will become eligible for Medicare.

A person is eligible for COBRA if her employer has 20 or more employees. In addition, the employee needs to pay the full policy premium and enroll within 60 days of leaving work. In order to obtain the additional 11 months of COBRA coverage, the employee must do three things. First, the person must be disabled during the first 60 days of COBRA. For example, a person who stops work due to her health on June 1 and applies for COBRA on June 10 will meet this requirement. However, a person who stops work, goes on COBRA, and starts another job will not be eligible for the 11 month extension, if she becomes unable to work later due to her health. This is because the person has a date of disability which is more than 60 days after the person started COBRA coverage. Second, the person must obtain an award letter from SSA during the first 18 months of COBRA. This means the person should make sure that the application for benefits is done correctly so that the application is approved within 18 months of filing. Third, once an SSA award letter is received the person has 60 days to notify the employer and insurance company that an SSA award letter has been received and to supply the award letter to the employer and insurer. If one of these three steps is not met, the person will not be eligible for the 11 month extension of COBRA.

3. I received SSI and Medicaid during the first five months that I was unable to work. Now I receive SSDI and my Medicaid coverage has stopped. Is this correct?

If a person gets a Social Security Disability Insurance (SSDI) check that is more than the state Supplemental Security Income (SSI) amount (typically $674 a month) that person loses SSI and cannot get

Medicaid in most states. For example, a person has an SSDI check of $1200 a month. An SSDI check of $1200 a month is more than the SSI amount of $674 so the person is not eligible for Medicaid in most states. A small number of states allow people with a low SSDI check to spend down a certain amount of money on medical expenses each month and then the state allows the person to receive Medicaid for that month. In almost all states, a person over the age of 19 years of age cannot be eligible for Medicaid unless he is also receiving SSI benefits or meets a Medicaid spend down. The Affordable Care Act, also known as Healthcare Reform, will make it possible for anyone, including adults, to obtain Medicaid benefits if the person's household meets a certain low income amount. This section of the Affordable Care Act does not go into effect until January 2014.

4. When I apply for SSA benefits should I apply for both SSI and SSDI?

A person needs to file only one application with the Social Security Administration (SSA). SSA will determine if a person is eligible for Supplemental Security Income (SSI), Social Security Disability Insurance (SSDI), or if the person is eligible for both programs. A person is eligible for both programs when the person has worked for a short time and so is eligible for a small amount of SSDI. When the SSDI amount is less than the SSI amount in the state, the person is eligible for SSI and SSDI. For example, an SSDI check of $200 would allow a person approved for SSA benefits to also receive an SSI check of $464, in a state whose SSI amount is $674. The good thing about an adult receiving SSI is that it allows the recipient to be eligible for Medicaid benefits. A person cannot "waive" SSDI eligibility so that the person would be eligible for SSI and Medicaid, even if the person has no other option for insurance coverage. ▲

Beth is 44 and has CF. She is a Director of USACFA and is the Secretary. Her contact information is on page 2. She is an attorney who specializes in disability claims. You may send CF-related questions of a legal nature to: bsufian@usacfa.org.

The Power Of Two – Premieres Summer 2011

THE POWER OF TWO
ザ・パワー・オブ・ツー

"The Power of Two" is a documentary film about cystic fibrosis (CF) and organ transplantation. It offers an intimate portrayal of the bond between half-Japanese twin sisters Anabel Stenzel and Isabel Stenzel Byrnes, their lifelong battle with CF, survival through miraculous bilateral lung transplants, and improbable emergence as authors, athletes and global advocates for organ donation. The film was directed by Academy Award nominated producer, Marc Smolowitz ("The Weather Underground" and "Trembling Before G-d").

Featuring expert interviews, archival footage and deeply personal testimony from the twins and others whose lives have been impacted by cystic fibrosis and organ transplantation from the U.S. and Japan, the film provides unprecedented insight into the personal, cultural and societal aspects of the illness journey and the modern medical miracle of transplantation. Without question, "The Power of Two" provides hope and inspiration through powerful personal stories of patients. This film will educate and enlighten viewers about living with chronic illness, patient advocacy, and cultural influences on health care access.

Watch the trailer for the movie here: http://www.thepoweroftwomovie.com/watch-the-trailer/.

"The Power of Two", in partnership with the United States Adult Cystic Fibrosis Association (USACFA), is proud to announce the release of this feature documentary film in five cities across the United States – San Francisco, CA (Sept. 10); Richmond, VA (Sept.13); Washington, DC (Sept.15); New York, NY(8/26-9/1); and Los Angeles, CA (8/16-8/22). Be among the first to experience the feature documentary film on the big screen in these major US cities! Check out the website for additional Fall/Winter screenings across the United States: www.thepoweroftwomovie.com.

Please spread the word to your friends, colleagues and community about this unique opportunity to see this one-of-a-kind, life-affirming film in these locations! We hope to see you there!

SPIRIT MEDICINE

A Receiving Spirit

By Isabel Stenzel Byrnes

Treasure the love you receive above all. It will survive long after your gold and good health have vanished. — Og Mandino

In this Spirit Medicine, I'm going to muse about the spirituality of receiving and how to cultivate a gracious way of receiving. By "receiving", I mean the receiving of gifts, blessings, help, attention – receiving just about anything.

Recently, my friend from Japan came to America with some savings in her bank account and a passion and ambition to receive a kidney transplant in America. Since there are hardly any transplants performed in Japan, she decided to pay out of pocket for the medical expenses in America. She has no family support and limited friends in America. All she had, really, was an openness to receive.

My friend was receptive to compassion. She allowed herself to be welcomed into homes, instructed on how to survive in America, to be fed, to be prayed for, to be driven to the hospital, to obtain care at a free clinic, to receive financial donations for her transplant.

I feel great admiration for my friend. I wonder if I could've done what she did, as it is very hard for me to receive. Yet, I've learned over the years that it's good for our spirits to receive. The Universe provides us with blessings and it's up to us to open our arms to them. Yet, sometimes, it's tough to welcome handouts.

I wonder if having CF plays a role in making it hard to receive things. I know a number of CF adults like me who yearn to be self-sufficient and independent. To receive help or assistance counters the staunch independence that we yearn for. Our American culture of independence and self-sufficiency implies that to rely on others, to need, to ask – to receive – is un-American. I fear that I might be smothering, annoying, resented. No one wants to be a leech.

Yet, the truth is, having CF forces us to rely on others to receive more than the average person, whether it's public assistance, rides to the hospital, help with cooking and cleaning, or even help getting up from a chair after a lung transplant. The list of our needs goes on and on - special medications, special nutritional supplements, even special medical devices. We have to keep on receiving if we are to keep on living. This is especially true for me as a transplanted person. I am called a lung 'recipient'. My very title defines my relation to another person – the giver or donor.

> *"I think it's healthier, spiritually and emotionally, to look at receiving something as a cycle of give and take."*

ISA STENZEL BYRNES

Even in a non-material sense, having CF invites us to receive special attention, special recognition as brave or inspiring. For example, this past May was the time of year when we have sent out our Great Strides solicitations or Mother's Day Tea letters, and plead to our friends and family for a donation. When the notices come in the mail or via email, listing who has donated, I am so touched. But I also find myself lowering my head, holding an embarrassed smile, and feeling awkward about receiving donations for myself and my disease.

So, why is it hard to receive?

Social manners dictate our behavior. It is socially polite to humbly and gratefully deny ourselves when we receive a gift or favor. All of us have, at one time or another, been given a large gift, and responded impulsively, "Oh, thank you so much, but I can't take this. It's too much. I can't accept your gift."

Receiving the generosity of others

can leave a host of emotions. I feel vulnerable, guilty, unworthy, or ashamed because I could never reciprocate such a gift in return. I can't reciprocate because my time, energy or money may be limited, especially if CF plays a role. I feel resentful if I feel the giver is trying to show off by giving such a big gift. I also don't want any gifts out of pity or sympathy. A repayment is in order. And, to be brutally honest, I don't want to feel forced or obliged to be 'nice' to that person just because he or she has been generous to me (I'd rather be authentically nice). This can be the case when we don't know or trust the giver.

Scholar and Rabbi Naftali Silberberg says, "To many people, giving is much easier, and more satisfying, than receiving. The act of giving allows the benefactor to feel important, valuable and productive. Giving is also the ultimate expression of one's humanness, the ability to transcend one's own needs and care for another. And even on a selfish level, giving earns the giver respect and admiration." I couldn't agree more; perhaps that is why I enjoy giving so much – to give offers a sense of meaning and purpose.

Sometimes our upbringing influences how easy it is to receive help. When I was a kid, and in and out of the hospital, my parents deliberately did not bring gifts for me to the hospital. They didn't want me to associate being sick with deserving stuff. They worried I'd become spoiled. I envied all my roommates who were lavished with flowers, balloons and teddy bears!

Much like a child can learn to receive with a sense of entitlement, it is also possible for us to learn to receive in a gracious way. It is a practice that takes time to cultivate. I think it's healthier, spiritually and emotionally, to look at receiving something as a cycle of give and take. When you take (or receive), then the giver is receiving the opportunity to do good for someone, and that gives him or her a positive feeling. That might be the basis of the Buddhist understanding of karma - that if you receive, you once gave, and if you give, you will receive again. This is a theme of what makes us human: interdependency.

Yet one cannot block the flow of karmic rewards. To honor the generosity of others, and to allow your heart to just rest in love and be cared for, is beautiful. It is about honoring the love that is extended to you through support. It is part of our spiritual health. Whatever I am given also gives me an opportunity to practice cultivating this gracious acceptance. The giver benefits from being generous, by feeling happy to see us receive a blessing. Generosity is an act of his or her unconditional love.

As I approach my fortieth birthday, I am trying to listen to the universe when gifts and blessings are presented to me, and simply accept them with openness and gratitude. The Bible says, "As God's fellow-workers we urge you not to receive God's grace in vain." (2 Cor 6:1) I'm trying to look at other people's gifts as love, even as God's love. They are passing on love through generosity, and to dismiss that would be offensive to them, to the Universe, to God. David Spangler in his book, *Blessing: The Art and the Practice*, encourages all of us to experience the rewards of being a conduit for God's blessings. All around us are opportunities to witness what we receive on a daily basis. We just have to pay attention to where and when we are being blessed. And paying attention to me also means feeling full of thankfulness. Gratitude makes receiving richer and more beautiful for all. To thank the giver, to thank the Universe, to thank God are ways that we can graciously accept what we've received. Even when our bodies fail, feeling and expressing gratitude is a magical way of attracting people's gifts and blessings at our time of need.

My Japanese friend's predicament has made me aware of the importance of accepting our blessings. After receiving so much from strangers, she is now recovering from a successful kidney transplant- after 11 years of dialysis. She created her own fate by being open to assistance from others. She understands interdependency.

If you have ever been like me, and struggled with being a recipient of tremendous support, care, help and service, I hope this article speaks to you. Next time a blessing is offered to you, I also hope you will be moved to receive it with acceptance, love and gratitude. ▲

Isabel Stenzel Byrnes is 39 and has CF. She lives in Redwood City, California. You may contact her at: Isabel@usacfa.org.

Mailbox

I just fixed my computer today and finally upgraded my home internet service and this [cfroundtable.com] is the first site I googled! I am so proud of how neat and organized it is. I am very excited to be able to see *CF Roundtable* on line and I enjoy it. Thank you for all your hard work.

Received via Website contact form
Bracha Witonsky
Brooklyn, NY

SPEEDING PAST 50...

If Only What I Know Now Had Been Available Then

By Kathy Russell

At long last it looks as if we may get some spring weather here in the northwest. Wait a minute, it's summer! Oh, well, we'll take nice weather whenever we can get it. I made it through another winter and spring without getting ill. I call that a real victory. As I get older, it seems that each spell of cooler weather is harder on me. I have learned that it is best if I stay indoors through most of the cold weather. Even with staying indoors, I still got pneumonia in both lungs, at the same time, last year. Such is life. All we can do is try our best to avoid the things that can compromise our health and handle whatever occurs.

One thing that I know now, that I may not have understood when I was younger, is that no matter how much I try to do everything right I still may get ill. I can be totally compliant and vigilant and still catch some nasty little bug or have my lungs fill up with some kind of gunk. At least, if I have been as compliant as possible, I know that I don't have to beat myself up for getting ill. It wasn't my fault. But, wait! Is it usually our fault for getting ill? I doubt it. I think that is just the way life is, when you have CF.

How I wish that I had started irrigating my sinuses when I was much younger. I might have avoided some sinus surgeries and almost surely would have avoided a lot of pain. Back when I was young, my ENT doc wanted me to use a bowl filled with warm salt water and to "sniff" this solution into my sinuses. I tried it, but my "sniffer" was so poorly coordinated that I always aspirated [inhaled] some of the solution and ended up choking. So I gave it a pass.

When I eventually had sinus surgery, I still didn't want to try sniffing salt water, post-operatively. After my third sinus surgery, my doc suggested that I try using a WaterPik appliance for irrigating. He explained that he modified the WaterPik tips and showed me how to use it.

We discussed what kind of solution I could use. He suggested that I use tap water and table salt. I declined, because of the anti-caking additives in table salt and the possibility of contamination of water taps. I said that I might try using sterile saline. We discussed the pros and cons of sterile saline. The only con that I remember is the price, but since my insurance would cover the cost, that wasn't such a big con. The pros included the fact that there would be no contamination of the salt or the water in a sterile solution.

KATHY RUSSELL

As we talked about irrigation, I asked about using either some sort of antibacterial solution or an antibiotic solution to help keep down the "bugs" in my sinuses. He had no suggestions, but I had one. My thought was that since we used acetic acid (vinegar) to keep down certain organisms, perhaps it might help my sinuses. We discussed the pros and cons of using such a caustic solution on tender tissues.

The doc wondered about using regular vinegar. I countered that vinegar is fermented and would have the potential to cause problems because of that. I suggested that using sterile glacial acetic acid (GAA), which is a pure chemical that is not fermented, might be a better choice. He did some research and found that there is a type of GAA that is used for ear drops, with some good success.

My doctor checked with the hospital pharmacy and found that they would compound a solution for me. It is a very low concentrate and I use only a little bit of it in my irrigating solution, but it works. I have been irrigating my sinuses for 16 years. During that time I have had no recurrence of polyps and have had only a couple of minor sinus infections. Getting rid of sinus pain was a real improvement in my life.

I wish that I had insisted that my doctors do something about my twisting spine, when it first started to twist. I remember the docs discounting my complaints and I felt that the tacit comment was that I "wouldn't live long enough for it to make any difference". Well, I fooled them and I lived. My spine has kept on twisting and causing discomfort. I think that I might be a little taller, if my spine weren't so twisted,

and that would be nice.

Between the twisting and the arthritis, I am getting shorter all the time. I have lost a few inches in height and I didn't have that much to spare! Compressed disks, compression fractures and scoliosis all have conspired to make me one short, twisted, old lady. Such is life. At least I am still here.

Good shoes that give adequate support are another important part of a comfortable life. I had been experiencing intermittent pain in my right foot. It would catch me unawares, when I stepped on it, and I would nearly fall. It could happen with any step that I took, so there was no way to avoid the pain. I talked with my primary care doctor about the pain and he referred me to a podiatrist [foot specialist]. That doc made casts of my feet and had custom orthotics [shoe inserts] made. What a difference they have made. No more pain and no more near falls. Because the orthotics are rather bulky, I don't wear a lot of stylish shoes. (And I like shoes!) It is more important to me to have no pain than it is to be stylish. I can move my orthotics between several pairs of shoes that will accommodate them, though, so all is not entirely lost. Making accommodations for our health becomes second nature.

One accommodation that I was making was unnecessary, it turns out. When I started using oxygen 24/7, I was experiencing irritation inside my nose. The ends of the "horns" on the cannula would cause sores in either the septum or the outer walls of my nose. These got to be quite unpleasant. I tried various methods to alleviate the problem. One thing that I did was to cut off the ends of the horns so that they didn't touch the sides of my nose. The only problem with doing that was that the ends were a little sharp and that could cause a whole new set of problems. I used some prescription cream on the inside of my nose, to try to keep everything healthy, but I still wasn't satisfied.

One day I went to my local hospital and asked at Respiratory Therapy if I could buy a pediatric cannula to try. I explained what my dilemma was and they were happy to let me give the smaller cannula a try. Wonder of wonders, it worked! The smaller cannula sits in my nose without bothering either the septum or the walls of the nose. That is all I use now. How I wish I had known that sooner.

As with many people who have CF, I have various gastrointestinal (GI) problems. I have suffered with GERD (gastro esophageal reflux disease) for as much of my life as I can remember. The terrible pain would wake me from a sound sleep. Although we tried many different things to ease it, nothing really worked. That is, until I tried proton pump inhibitors (PPIs). The first one that worked for me was Prevacid. When my insurance provider insisted that I switch to Nexium, I gave it a try. It turns out that Nexium works well for me, too. Hooray! Another problem solved. If only Nexium had been around many years earlier.

I have had surgeries to repair various troubles in my GI tract. I have to wonder if I might have been able to avoid some of those troubles, if I had known about Mira Lax long ago. Many people who have CF experience GI trouble because of low or not enough digestive enzymes. I have had my years of taking supplemental enzymes, but I also have had many years where I didn't need to take them.

After many years of having the typical CF problems with frequent, bulky, foul-smelling stools, my body decided that it would prefer to be constipated. When I asked the CF GI doctor about this, she said to take more enzymes. She said that it was impossible to take too many, so just keep adding more until the problem is fixed. (Of course, we now know that wasn't quite correct.) So I added more enzymes and eventually had trouble because of it. (I have had half of my colon removed.)

When I talked with the late Dr. Jack Jacoby about my discomfort, he told me that many of his patients took bisacodyl (the generic form of Dulcolax) every day. After we discussed it some more, I decided to give it a try. It worked fairly well for a time, but after a while, it didn't work as well. Sometimes I would have to take double or triple my normal dose in order to get relief. It still wasn't ideal.

Last November, I ended up in the hospital with a twisted small intestine. Not pleasant but not too awful, either. I had nothing by mouth, other than ice chips, and had a naso-gastric (NG) tube in place for a week. After the intestine straightened itself out, they started to let me take things by mouth. They wanted to get everything moving again, and one of the meds that they tried was Mira Lax. Wow! It worked easily and quickly. My intestinal discomfort was gone! I couldn't believe it.

After I was home and back to my normal routine, I asked my doc if I could try Mira Lax instead of bisacodyl. He was okay with it, so I made the change. I still am happy with it. I take a slightly smaller dose than what is recommended and am very happy with how it is working. If only this had been around and I had known about it many years ago.

As you can see, there are so many things that are available to people who have CF now, that weren't available when I was young. I hope that these and other things will help to make your life more pleasant and even longer than mine.

Stay healthy and happy. ▲

Kathy is 67 and has CF. She is a Director of USACFA. Her contact information is on page 2.

The Blooming Rose Foundation

By Kathleen East

I entered the world of CF five years ago with the birth of my beautiful daughter, Maylie. At the time, CF screening was not mandatory and since I was unaware of the disease, I did not request the test. At birth, Maylie appeared healthy. She weighed over eight pounds, but she was not an easy baby. There was no amount of consoling that would soothe her screams, and she seemed to be constantly in pain. I look back at it now and I'm filled with regret. A war was raging in my daughter's body and I didn't even know it.

At six months, Maylie weighed a healthy 22 pounds, but she had developed a chronic cough due to aspirating pool water. Her bowel movements became frequent and plentiful, and soon overwhelmed my cloth diaper regimen. She would exhaust an entire bag of Seventh Generation diapers every day. I didn't know what to do. I somehow convinced myself that this was just the way her body functioned.

After six months and a round of Zithromax, Maylie did not display additional CF symptoms for nearly a year. But, by the time she was two, she aspirated chlorinated water again and the coughing and GI symptoms worsened. We went to our family practitioner. It became a weekly routine, but there were no answers–there was only helplessness. After a confusing X-ray, we were referred to a pulmonologist. A month later, Maylie was diagnosed with CF.

By this time, Maylie had pneumonia, her oxygen saturation hovered around 80%, her fingers and toes had begun to club and she had gained only one pound in the last two and a half years.

When we received Maylie's sweat chloride level of 138, the pulmonologist, who was on his way to the European CF conference later that day, came in for a brief discussion about CF. Fifteen minutes of his time was all we were graced with. I clearly remember his words. He said my life would be hard, that it would be devastating at times but, in a couple of years, it would become my reality. I would know more about CF than most doctors. Then he said, "Sorry I don't have any handouts for you, but you can get those at the CF clinic in April." April was two months away. I was alone with this

KATHLEEN EAST

diagnosis for two months. I had nowhere to turn; I knew enough to know that the Internet was too dangerous for such a search. So I waited.

The diagnosis was devastating, but I felt a touch of relief. For the first time I could see beyond the questions and the fear. Now there was a reason for my daughter's exhaustion, her fragility and her misery. Now there could be a plan. Now we could move forward and Maylie could start living.

When faced with a diagnosis like CF, you quickly learn to fight. My ring was advocacy—not only for Maylie, but for everyone sharing her journey. Educated families, single moms, young parents, individuals with or without support systems–we all need help. Because of this, I started the Blooming Rose Foundation (BRF) to offer support, education and advocacy services to individuals and families living with CF. Our primary program–Newly Diagnosed–offers families a package of pertinent information about CF, patient assistance programs, samples, drug co-pay cards and links to CF blogs, Websites, non-profits and social networks.

After working in the CF community for a year, I became aware of the large population of adults who live and thrive with the disease and I wanted to ensure that BRF addressed each stage of CF. I also realized that not everyone has correspondence with a CF team between appointments and that many individuals do not feel comfortable questioning their team. Without a continuous discussion of fresh options, it is easy to wallow in treatment ruts. In an effort to address these realities, BRF formed a panel of experts to answer questions pertaining to CF.

The BRF panel strives to fill these gaps and provides information and inspiration to individuals and families with CF. It blends the expertise of medical professionals and the passion and experience of individuals, advocates and parents living with CF.

Of all my interactions as a social worker, I have never witnessed a group of individuals and organizations as inspiring as the CF community. Of course I, like everyone else, wish for a control for my daughter and my friends, but it is the CF community that provides daily motivation. I am honored to walk among the heroes and warriors that Maylie's disease has introduced to me and it gives me peace to know that these inspiring individuals will be Maylie's peers for the rest of her beautiful days. ▲

Kathleen is the founder and CEO of the Blooming Rose Foundation. She is the mother of two amazing little girls. They reside in Bozeman, MT. You may contact her at: kat@brfcf.org

DESCH *continued from page 1*

and need to get to the lab for blood draws three times each week. Did I mention the port dressing changes? I literally needed a flow chart to traverse through my day, or I would forget an essential item. Of course, most people get admitted to the hospital for this rather intensive treatment time, where, at least in theory, they keep track of these things – wink – but I hate hospitals, and go in only if it is absolutely mandatory.

Enter Tonic. As luck would have it, I was able to beta test Tonic this "winter from hell". Tonic is an app that helps you to remember and track whatever you need to track. Setting up Tonic is a simple task. You simply make a list of all that you want to remember and track. It took me about 30 minutes, because my list of "tonics" was enormous. When it was done, though, my trusty little iPad would bleep whenever I had to do something. Take out med (bleep). Infuse this (bleep). Take this (bleep). Eat (bleep). Weigh yourself (bleep). Eat more (bleep). Trust me, the thing was bleeping all day, every day, of each exacerbation. But I was so completely on track that I missed nothing! Nor did I do or take anything more than I was supposed to, because you check off each "tonic" as you do or take it. This is a very satisfying property of Tonic for anal compulsive people who like checking things off lists (as I do).

Second, remember that the idea is to make your life easier, not to add another chore to your day. When my life is so complicated that I really need help, I use Tonic. Other times (like now) when things are pretty good, I let Tonic fade somewhat into the background. It still reminds me, but I turn off the "bleep" alarm and don't always check off all the activities or enter a lot of data. You will find your own balance.

In summary, Tonic is just what I needed at just the right time. I would recommend it to anyone, whether you have a complicated regimen like mine, or just want to track a few things. If you decide to give Tonic a try, let me give you two hints. First, use it for all of the things you do for your health – everything - really. Meds, sleep, exercise, nutrition, water intake, everything you can think of that you know you want to do to be at the top of your game. For instance, Tonic even reminds me to journal, to stretch my hamstring muscles, and to meditate.

You can find Tonic on the web at: http://www.tonicselfcare.com. ▲

Julie is 50 and has CF. She is the President and Founder of New Day Wellness. http://newdaywell.org.

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Announcements

VOLUNTEERS NEEDED FOR STUDIES AT NIH

The Pulmonary-Critical Care Medicine Branch of the Department of Health & Human Services, National Institutes of Health(NIH), National Heart, Lung, and Blood Institute, in Bethesda, Maryland is conducting **a research study to evaluate the role of bacterial products involved in lung disease in cystic fibrosis.** We are looking for individuals with cystic fibrosis and Pseudomonas aeruginosa. The participants will be seen at the NIH. They will have blood drawn (around 2 tablespoons) and also have a sputum sample collected. The participants with CF will be paid $50.00 for taking part in this study. We will pay for the transportation of patients who do not live in the local area. If you have CF, are at least 18 years old, have Pseudomonas aeruginosa and are interested in more information about this study, please call us collect at (301) 496-3632 or send E-mail to: barnesp@nih.gov.

A research study of hereditary factors associated with cystic fibrosis and other lung diseases is being conducted at the Department of Health & Human Services, National Institutes of Health (NIH), National Heart, Lung, and Blood Institute in Bethesda, Maryland. Participants will be admitted for an overnight stay at the NIH to have blood drawn, a PFT, chest x-rays, and EKG. Assistance with travel costs as well as a $150 stipend will be provided. If you have CF, are 18 years of age or older, and are interested in participating in this study, please call us collect at (301) 496-3632, or send E-mail to: barnesp@nih.gov.

We are looking for individuals with cystic fibrosis who previously participated in NIH studies. If you have taken part in an NIH study, please call the toll free number: 1-877-644-5864 and select #3 on the menu; or send an E-mail to: barnesp@nih.gov.

FOCUS TOPIC

IF ONLY I HAD KNOWN THEN WHAT I KNOW NOW

Learning to Manage My Health

By Debra Radler

Cystic fibrosis, the grand master of mucus, is a disease that requires managing. There is no way around it. And the one thing that I do know now that I did not know then is that without proper management, my health would be in the tank faster than I could spell N-E-B-U-L-I-Z-E-R! Over the years I've learned how to better manage it, and how to adapt to its ever-changing hues. I will attempt to share with you some tried-and-true tricks that work for me, as well as some philosophies that I've developed along the way.

Savor life. Do not, under any circumstances, believe all that you read about this disease regarding life expectancy. And do not allow yourself to prematurely fret over the imminent decline in your health before your health actually does start to decline. You can waste precious years anticipating the end without enjoying the beginning. I did just that as a child, and I wish that I had known better.

Exercise. Do it as often as you can. Do your best to make it a discipline rather than something you dabble in. It will add years to your life.

Hygiene. Brush your tongue with a damp toothbrush covered with kosher salt. It does wonders for the bacteria build-up that is inevitable, given what we cough up daily. I tried every method of oral hygiene and nothing has worked as miraculously as brushing my tongue with salt. It makes it perfectly pink.

Mucus expulsion. It's a gross topic, yes, but a daily part of most of our lives. Consider using a spit tureen, such as an old margarine bowl, instead of tissues, during therapy. I find that I am better able to monitor the volume and viscosity of my mucus secretions, and I think that it is more hygienic to rinse and dump it down the toilet than to accumulate hundreds of germ-infested tissues in my house.

Photograph by Michael Slaughter

DEBRA RADLER

Toilet paper vs. tissues. I like using toilet paper instead of boxed tissue. I like the softness of the roll and I keep it next to my pillow at night for easy access.

Sinus rinsing. I have Kathy Russell to thank for turning me on to this treatment a few years ago in one of her articles. I cannot stress the benefits enough. I use the Neil Med Sinurinse kit that can be purchased from any drug store, and I rinse my sinuses at least twice a day and more often if I feel a cold coming on or if I'm exposed to allergens. In addition to the salt water rinsing, I also rinse once daily with 8 oz. of warm Sterile Saline Solution and 4 eye droplets of prescription Acetic Acid….the purest form of vinegar. It is an amazingly effective cleanser. That which does not belong in the sinuses will be flushed out with regular practice of this therapy. It helps to keep the germs from passing from sinuses to lungs. It is a practice that has changed my life, and I wish I had known about it earlier.

Chest PT as one option, not THE option. Isometric breathing exercises use two accordion-like drums, which are squeezed together to promote resistance while we contract our muscles and exhale slow, controlled, long breaths. It is a learned discipline that was demonstrated in Chicago years ago at one of the CF Awareness Day conferences. I was sold immediately when I saw the amount of productive coughing going on. I became even more sold when I participated in a study to measure the benefits of this practice. It was my chosen form of therapy for years.

The Therapy Vest seems to work for me only when I incorporate deep breathing exercises, muscle contractions, arm flapping or upper body rotation in my therapy session. If I just sit still and shake while watching television or reading, I will not have a productive session. I have to put in a little more effort in the therapy to make it work for me.

Port placement. Decide where you ultimately want your port placed–arm or chest–and advocate to have it placed exactly where you want it. I wanted it placed in the upper arm, and the interventional

radiologist denied me that wish and placed it in the lower arm. It is an unsightly addition to my body. I have since learned that upper arm placement is relatively common and another interventional radiologist confirmed to me that he would have placed mine in my upper arm. I know now, and did not know then, that I could shop around and self-advocate and find the right radiologist for my needs. I wish I had known.

Nebulizing. I boil my nebulizer cups a couple times a week. I rinse and disassemble each cup after each use, but I accumulate the used cups in a large lingerie bag until I am running low. I then submerge the entire bag into a large pot of boiling water and boil for 9 minutes. When done, I remove the entire bag with a wooden spoon that I twirl around the bag in order to grab it, and I place it on a mound of paper towels to dry. I have found this to be the most time-saving way of cleaning my equipment.

Potty training. Who among us has not had to navigate the dance between intestinal blockage and intestinal gushing? It's not pretty and it can enslave us to the toilet. When I find that things are just not moving along as they should, I use an electric percussor and place it on the lower abdomen to vibrate the intestines and get things moving along. It actually works so well that I have been known to bring it on vacation with me, despite the additional weight in the luggage.

Time management. Realize that with the progression of disease the amount of time devoted to it increases substantially. It's important to have some systems in place so that the day does not end up being entirely devoted to cystic fibrosis. I find that incorporating health promoting activities is a great way to service the needs of the disease and provide an outlet for enjoyment as well.

This was just a sampling of the things that I've incorporated into my life to better manage my care. And without the generosity of others in the CF community sharing their knowledge with me, I might not know about half of these things. When I look back to how little I knew about caring for this disease in my teens and early twenties, I'm amazed by how far I've come. At forty-nine I feel that I know quite a bit. It will be interesting to see how little I do know, when I look back from the perspective of a 60 year old!! ▲

Debra is 49 and has CF. She is a semi-retired CPA who lives with her husband, Adrian Gulinski, and two Bichon pups in Roselle, IL. She is a proud stepmother to David and Nicole. She can be contacted at: debraradler@hotmail.com

New Online Nomination Form!

http://www.heroesofhope.com/heroesofhope/nominate

The Heroes of Hope™ Living with CF program is delighted to share with the CF community a new Heroes of Hope online nomination form. Now nominators can easily nominate the CF heroes in their life with this handy, online submission form. The form can be filled out at http://bit.ly/dQx6C4. We all know heroes in the CF community who are deserving of this award, so nominate someone today!

"We are thrilled to launch our new online nomination process for the Heroes of Hope Living with CF Program," said Heroes of Hope panel member and CF advocate Anabel Stenzel. "We hope this makes it easier for people to nominate their favorite CF hero. We look forward to receiving many more nominations so we can celebrate and recognize the gifts of many worthy people living well with cystic fibrosis."

Heroes of Hope are individuals who serve as role models and portray hope to others with CF while proactively maintaining their health. They are motivated to live life to the fullest and do not let the limitations of CF get in their way of pursuing dreams. If you know someone with CF who fits this profile, we encourage you to submit an online application form today by clicking here!

Heroes of Hope™ Living with CF, sponsored by Genentech, recognizes individuals with cystic fibrosis (CF) who live full and productive lives while managing their healthcare needs. To date, Heroes of Hope has honored over 70 outstanding individuals with CF around the United States. Heroes of Hope recipients inspire others through their outstanding accomplishments, dedication to health maintenance and contributions to their communities. To learn more about the program, go to www.HeroesofHope.com.

FOCUS TOPIC

IF ONLY I HAD KNOWN THEN WHAT I KNOW NOW

The View From The 9th Floor... For Cathy

By Laura Mentch

The building loomed so large… not its size, but the thought of it. For the past few years my nurse advocates for a hospital tune-up while I bargain with her to stay home. I believe that I can take care of everything: my work, my home, my family and my health. I've not ever been in the hospital for CF and her suggestion to go there is discouraging.

Eight years ago I jumped into the CF world with a crash course in nebs, vests, PICCs and all the meds. After my first clinic visit I added it all into my busy life: rushing morning treatments to get to work; teaching and working through IV infusions while making every meeting and family event – keeping all of the plates spinning. When I am really sick, I thought, that's when I'll need to go to the hospital. The reality is that I haven't been really sick since I was diagnosed and first walked into the CF clinic. It's hard to grasp that a hospital stay could be routine.

LAURA MENTCH

This year spring arrives late. The change in weather and a sneezing, runny nose usher in an exacerbation. Although I was just there, I return to clinic. Following my weak protest, a plan is made to admit me. Soon I am on my way to the hospital. With hesitation, I step out of the elevator and cross this threshold, settling into my room on the 9th floor.

Soon I am engaged in the rhythm and routine here. I am up early, the sky brightens, cars fill the roof of the parking garage, the nurse starts the IV and there are noises outside my door as the floor awakens. Breakfast appears as the respiratory therapist sets up; later the CF team arrives. I stop negotiating to leave in a few days – I'll be here for Mother's Day. The respiratory therapists teach me a lot, as my nurse expected, and I learn why some call this a clean-out. I think how many ways there are to cough, like a hundred words for snow. So many people are helping me and coordinating my care. I cannot get support like this where I live, 700 miles away. After a few days looking through this

Changes

By Laura Tillman, President USACFA, Inc.

As the seasons change, so do things here at USACFA. We would like to thank Anne Williman for her tenure on our board of directors and wish her well in the continuing years. Her proofreading and editing have proven so invaluable that we have asked her to continue on in that capacity despite no longer being a director. So, we're not really saying, "Goodbye" - just a heartfelt thanks for her time serving on the board!

Additionally, we would like to welcome Mark Levine to our board of directors. Mark hales from Michigan, where he lives with his wife and two children. He brings a great degree of enthusiasm and experience to his position as director and we are very pleased to have Mark join us. Please be sure to read his biography on page 15.

Other changes that have occurred or are occurring include a newly designed Website, a Facebook page, blogs and Twitters! You may now subscribe online and receive your newsletter either online, as a hard copy, or both. Be sure to take a look at us at: www.cfroundtable.com and to "Like" us on Facebook.

We hope you enjoy this issue. It brings a lot of new perspectives to how we've dealt with CF past and present!

window, my perspective about being in the hospital shifts. I'm surprised how much I like it here.

Days later, night approaches and I prepare to go home. The parking lot empties and I have another respiratory treatment as the floor quiets. Two hours until the last IV of the day. I lower the shades and reflect on this experience. My biggest lesson is understanding that this is the time to focus on CF. It is ok to set down some of the plates, learn to nap and let others help me take care of myself. My challenge is to follow through, especially when I am home.

What could have helped me see this earlier?

Leaving, I look over my shoulder at the building and wonder if I am going home too soon. I understand that I will be back and when that happens, I'd like to have a room on the other side of the floor so that I can watch the mountains. ▲

Laura is 58 and has CF. She and her husband, Michael Brody, live in Bozeman, MT. Her work in health education focuses on family planning and sexual health.

In Memory

Lori Kipp, 38
Pacifica, CA
March 26, 2011

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

Meet Our New Director

MARK LEVINE

Mark Levine, 41, is the newest board member of USACFA. Diagnosed with CF at the age of 2, he has worked with the Cystic Fibrosis Foundation for many years in four different states. Mark grew up in Connecticut with his brother, David, who also was born with CF, and their parents, Jane and Len. Mark attended Lehigh University in Bethlehem, PA, graduating with a BS and MS in Mechanical Engineering. It was in PA where Mark became more active in CF activities – joining a support group and volunteering for the Foundation. While Mark was in college, David, at the age of 18, received a lung transplant. He passed away just three years later after suffering from rejection. Immediately after finishing school, Mark moved to Michigan to start a career with Chrysler, where he has been employed for 17 years. A nine year relocation to Indiana to work in manufacturing introduced Mark to the Indiana Chapter of CFF where he became very active. He was a bachelor in a CF auction and later served as chairman of the auction, which is Indiana's largest CF fundraiser. He was a founder and chairman of the Kokomo, IN, Great Strides Walk and has spoken to dozens of audiences at CF fundraisers throughout Indiana and Michigan. Before moving back to Michigan in 2006, Mark met his wife, Joelle, in Indiana. With their two children, Brooke (16) and Adam (14), Mark and Joelle now reside in West Bloomfield, Michigan, where Mark also serves on the CF Foundation Board of Directors. Mark is excited to be a part of USACFA.

Mark is 41 and has CF. He is a Director of USACFA. His contact information is on page 2.

Pulling My Head Out Of The Sand

By Jennifer Eisenmann

If only I had known then what I know now. I hear this phrase from friends all the time referring to the stupid risks they took in their youth, the partying they wish they hadn't done and the bad relationships they put themselves through. But when you're talking about having regrets with how you took care of your health, it is a whole different ballpark. Although this article can apply to anyone, I think it is more for the teens and twenty-somethings who have CF, still feel pretty good and don't want to deal with consistently doing treatments.

First of all, let me just say – I was you. I was not a very sick child. Sure, I woke up earlier in the mornings so my mom could do CPT on me, I got colds more often than other kids, I had to take enzymes and I certainly had more stomach aches and digestive issues than my friends. But I was hospitalized only once for a blockage and once for an almost burst appendix in all of my youth. I consider myself very fortunate. The unfortunate part is that when I got into my teens, I saw no need to do any kind of airway clearance and my doctor at the time did not have me on any inhaled meds – not even albuterol!

I knew CF could be a serious disease but just thought I had such a mild case that I would be fine. I stuck my head in the sand. I went off to college with promises to my parents to "do my treatments", which at the time consisted of this ridiculous contraption that sort of went back and forth and hit my chest. I didn't see that it did any good and I "felt fine" so why bother with it? I knew my lung function was somewhere in the 90s percentile-wise (I knew nothing of FEV1, or what any of it meant – another part of sticking my head in the sand) and I figured it would stay that way for years.

My friends in college liked to do a lot of hiking and I started noticing that I would get out of breath more easily than they did, but I just chalked it up to my "mild" CF and considered it no big deal. It wasn't until I moved to Michigan, at age 22, and started having major coughing fits that I decided to find a CF clinic and get some real answers. And let me tell you, I was not prepared for what the doctor had to say. He said if I continued living the carefree life I was living, I might not make it to age 26. What???? Although this was a harsh thing to say and probably a little exaggerated, it was exactly what I needed to hear. I was immediately admitted to the hospital for my very first "clean out" and was put on albuterol and Pulmozyme. Soon after that the first Vest prototype was out and I began to use it for airway clearance.

JENNIFER EISENMANN

But I STILL left a little part of my head in the sand and wouldn't do more than one treatment a day. I had a life to live! I did not ask for this! One treatment a day was already getting in the way of my life – don't ask me to do two! And despite the cold hard facts displayed in front of me, I still had a little bit of that "invincibility syndrome" of youth. You know - that syndrome that seems to cause younger people to take wild risks that they wouldn't even consider when they get older?

It was a slow and tedious process to get myself to be a compliant patient. It took many clean-outs and many scared moments. And, of course, hindsight is 20-20. I am now 40 years old and have been compliant for over 10 years. But with my FEV1 in the 45 percent range and occasionally dipping down into the 30s, and arms that are unable to endure many more PICC lines, I truly believe I would be much healthier now and have a better quality of life if I had taken better care of myself when I was younger and "felt" healthy.

If I knew then what I know now, I would have done my treatments diligently, even when I felt good, to prevent my future self the hardships I endure now. ▲

Jennifer is 40 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Living Life with CF, Acceptance and Gratitude

By Bonnie Bleiweiss

I am an adult, newly diagnosed with CF. I was genetically identified six years ago at age 58, but because I also have Alpha One Antitrypsin Deficiency, a liver disease that has lung symptoms, my CF symptoms were ignored until two years ago. In the past two years I went through a myriad of lifestyle changes putting me off the stress test charts. We relocated from San Diego to Houston. I retired and went on Social Security. My husband's new job offer was withdrawn after we arrived, and along with it, our health insurance. I spent six hours a day on the web and the phone seeking assistance and insurance for three months, all the while not getting treatment.

I had a career in Commercial Casualty and Health Insurance and that helped me negotiate a plethora of websites and assistance organizations. Then I was admitted to the hospital for a blood clot at the insertion point of a new Port-a-Cath, which was implanted to receive augmentation therapy for the Alpha One. While in the hospital it was discovered that I needed oxygen while I sleep, so now I use it every night and when I exercise. I suspected all the time that the augmentation therapy wasn't really helping the symptoms I was having, so I had the port removed.

It was during the hospital stay that I was officially diagnosed with CF. I then found the CF center here in Houston and jumped into action by applying for and receiving Social Security Disability. Since receiving CF treatments and understanding the protocol, my symptoms are finally being addressed.

Additionally, to increase my stress level, my mother passed away in November of last year with non-CF related emphysema and heart problems. Due to the economy, my husband lost another job and we went on COBRA until he found work. (Cobra payments were more than our monthly rent.) Unfortunately, the economy and the cost of treatments and drugs led us to file bankruptcy.

Why do I tell you all these things? I'm sure I'm not the only person to have had these challenges. It's been difficult to be so alone and isolated at 64 years of age. No one in my family has CF, that we are aware of. I have no friends who have it, and being new to Houston, I had no friends. I am sad that I will never get to hug a fellow CF adult or really get to know them; but in spite of it, I'm overcoming the loneliness by joining the local YMCA, joining two local social service groups, and making peanut butter and jelly sandwiches for the homeless, offsite, away from the homeless themselves. I take Intermediate Yoga three days a week, with my oxygen tank in tow, and I walk as much as I can. It's fun to get a 'thumbs up' from passing walkers who see the oxygen back pack that I'm wearing.

Having CF has been an odd blessing and I live my life daily by acknowledging what I'm grateful for. CF gave me the excuse to spend $149 for a Kindle that I read at the clinic and during treatments. It gives me permission to slow down, meditate and enjoy my dogs. It has allowed me to maximize my multitasking abilities while creating craft-art and doing computer work; and it has given me the ability to laugh at myself when I talk to my dogs and hubby during 'vest' treatments. The dogs are truly perplexed! And lastly I'm grateful to CF *Roundtable* and the tireless contributors who share their stories and wisdom with all of us. ▲

BONNIE BLEIWEISS

"Having CF has been an odd blessing and I live my life daily by acknowledging what I'm grateful for."

Bonnie is 64 and has CF. She lives in Houston, TX.

If I Had Known Then What I Know Now

By Nichole Matthews

There are the typical rules for living with CF: don't go within three feet of another CF patient or sick person, do your treatments, take your medications, do this and make sure you do that. It gets tough sometimes and it's a constant hassle. But you know what? WE are here and WE are breathing!

When I was younger I had a tough time. In elementary school, between teachers not understanding about me being sick and my peers knowing I was sick and not wanting to be near me, I was a pretty lonely kid. I had my little sister and my cousins and that was about it. I looked forward to the Great Strides walk to see my other CF friends, because they knew what it was like to be lonely sometimes. Kids would make fun of me for my barreled back, my "weird" looking fingers, my constant cough and my continuous bathroom breaks. I wanted to be normal.

As I grew older I realized I was the stronger of my peers. I did not fuss over the little things in life and I cherished everything brought my way. I had a HUGE group of friends in high school and college. I was not shy about my disease. I told people right off the bat, "I have a genetic disease called cystic fibrosis. Please don't judge me. I am stronger because of it." Having CF isn't easy – one bit, but it is who I am. It made me who I am today. It helped mold me.

NICOLE MATTHEWS

If I had known back then what I know today, or even five years ago, I think I would have been a happier child. Life would have been a little easier for me. I might have been a child who didn't hide from people because of the laughing they did towards me. I would have tried harder in school and I am almost sure I would have had more friends, because of my outgoing attitude with my disease. I would have shared the information rather than shrugging it off. I would have spread the word of CF with others, to bring awareness to my community. CF is not something to be ashamed of but something to almost cherish, in a very twisted kind of way. I have these health problems for a reason. I am strong enough to live with them. I can try my hardest to overcome them and I will never give up. There were times when I wanted to give up because of CF, but with what I know today, I will not give up for quite some time. I am a strong woman who has conquered many things.

I am only 23 years old, but I am strong. I live every day with a sunny outlook, cystic fibrosis, gluten allergy, scoliosis, rheumatoid arthritis and heaven knows what else. But, I am here. I am living and breathing. And I am volunteering. Don't dwell on the negatives in life. Count your blessings. ▲

Nicole is 23 and has CF. She lives in Orchard Park, NY with her two dogs and three cats. You may contact her at: nmatthews@westherr.com or on Facebook.

NEW COLUMN

In the Spotlight

Many people have written to tell us how much they enjoyed our former column, *Unplugged*. *Unplugged* was a column where people could read about others dealing with CF. We soon will be replacing the old column with *In The Spotlight*. *In the Spotlight* will be similar to *Unplugged* with people answering questions and providing a window into their lives.

We will be interviewing people with CF and, sometimes, their caregivers, family members or medical staff. Currently we are looking for people who are interested in participating. *In the Spotlight* will be edited by Jeanie Hanley and Andrea Eisenman. You may contact them by email, Jeanie Hanley at: jhanley@usacfa.org or Andrea Eisenman at: aeisenman@usacfa.org.

If you want to participate, please e-mail us your contact information: e-mail address and phone number. We are looking forward to hearing from you!

PHOTO BY STEPHEN BOYER

Presence

The familiar presence of absence
Nearly listens before being comforted
By a chorus of warm and treasured
Embers of the heart in loving unison
Hovering over flickering votive candles
Representing angels unaware
And numbering one more loss
Than my heart seems capable of enduring.

Their lives and legacies
Form the beautiful tapestry
That I wear every day
As the familiar presence of absence
That I hear
In between distractions.

John Bartelt, 2002

"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...

JULIE DESCH AND HER IPAD WITH THE TONIC APPLICATION.

DEBRA RADLER AND HER HUSBAND, ADRIAN GULINSKI, ENJOYING FOUR YEARS OF MARITAL BLISS.

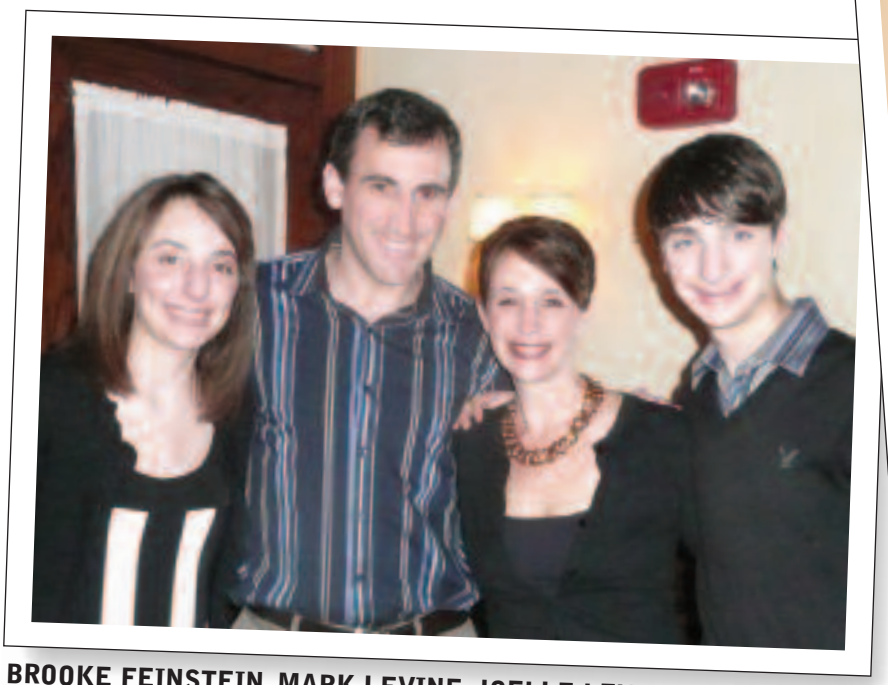
BONNIE AND SHELLY BLEIWEISS AT A RESTAURANT IN GALVESTON OVER MEMORIAL DAY WEEKEND.

WALTER BARTHOLOMEW WITH HIS CHILDREN, NICHOLAS AND VERONICA.

Photograph by Nicole Wickens, Green Door Photography

LAURA MENTCH (FOURTH FROM LEFT) AND FAMILY, MAY 2010.

KATHLEEN EAST AND HER DAUGHTER, MAYLIE.



BROOKE FEINSTEIN, MARK LEVINE, JOELLE LEVINE AND ADAM FEINSTEIN.

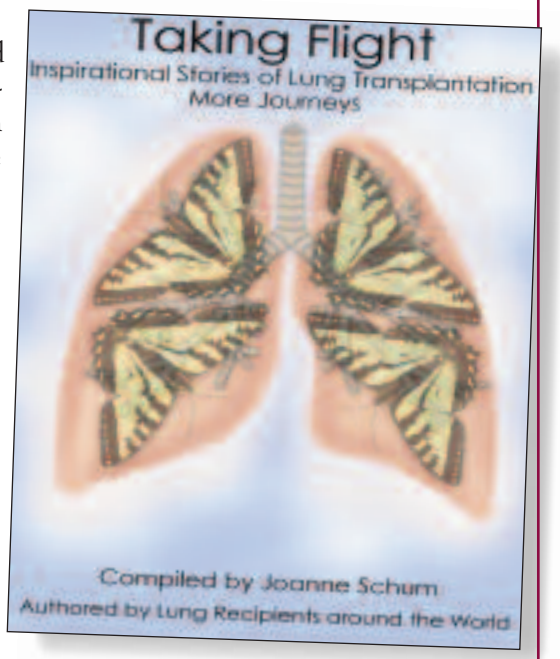
FROM LEFT: HUSBAND, JOHN; DAUGHTERS, MARIA AND JESSICA; NEW GRADUATE KEVIN; AND JEANIE HANLEY.

BOOK REVIEW

Taking Flight

Inspirational Stories of Lung Transplantation More Journeys

Compiled by Joanne Schum
Authored by Lung Transplant Recipients around the world
Reviewed by Kathy Russell



This book is a continuation of Joanne's earlier book and has more stories of transplant journeys. Most of the essays are written by people who have received lung transplants. There are a few essays that are written by friends or family members of transplant recipients and at least one by a member of a donor's family. All the essays talk of transplants from the writer's perspective.

This book is a good read for anyone who is contemplating lung transplant and for their families, friends and caregivers. It always is good for us to get a look at something from another person's angle. These stories come from many countries and allow us to learn about lung transplantation for reasons other than only CF. Some of the people have bronchiectasis, COPD/emphysema, pulmonary fibrosis or any one of several other conditions.

At a little over 300 pages, this book could be picked up and read right through. I doubt that anyone would do that however, because of the nature of the essays. It is possible to pick up this book and read an essay, then put it down. Since the stories are very touching, this may be the best way to do it. However you choose to read it, I am sure you will find it worth your time.

For more information about the book and how to purchase it as well as for information on when Joanne might be in your area, go to: Twoluckylungs@juno.com or luckylungsforjo@aol.com

Information from the Internet...

Compiled by Laura Tillman

This issue brings a potpourri of articles from the Internet

TREATMENTS

Aztreonam lysine for inhalation solution: a guide to its use in cystic fibrosis. Drugs & Therapy Perspectives: 1 April 2011. Volume 27, Issue 4 - pp 1-4

Aztreonam lysine for inhalation solution (AZLI) is a new option for the treatment of Pseudomonas aeruginosa lung infection in patients with cystic fibrosis. When compared with placebo in clinical trials, AZLI improved respiratory symptoms, pulmonary function and sputum P. aeruginosa density, delayed the time to need for inhaled or intravenous antipseudomonal antibacterials when used after a course of tobramycin inhalation solution, and was generally well tolerated.
http://tinyurl.com/46c2w7d

Disposable versus reusable jet nebulizers for cystic fibrosis treatment with tobramycin. Laurent Vecellio, Mohamed E. Abdelrahim, Jerome Montharu, Julien Galle, Patrice Diot, Jean-Christophe Dubus. Journal of Cystic Fibrosis. Volume 10, Issue 2, Pages 86-92

Jet nebulizers are commonly used to administer aerosolized tobramycin to CF patients. None of the disposable nebulizers tested in this study can be recommended as an alternative to the Pari LC Plus nebulizer for tobramycin.
http://tinyurl.com/5rhcd7r

Clinical use of dornase alfa is associated with a slower rate of FEV_1 decline in cystic fibrosis. Michael W. Konstan MD, Jeffrey S. Wagener MD,

Continued on page 27

Had I Only Known – Ugh Blood!

By Jeanie Hanley

Because I know that most of us with CF are fairly thick skinned, what with the coughing, secretions, digestive issues, etc., this article won't adversely affect most of you. All the others – you have been duly warned. I chose to write about hemoptysis – or coughing up blood. Not long ago I underwent a bronchial artery embolization to stop the hemoptysis. Under this procedure the suspected artery was hit with a medication that stopped the bleeding in its tracks. It's used only for those who have frequent, massive episodes of hemoptysis. Good times.

How my CF lungs progressed to that point is clear – repeated bouts of infections, mucus plugs, stress, allergic reactions and more - events that made my airways more fragile, eating away at the airway, causing bronchiectasis or ballooned out airways that do not function normally. Thus, effective cleaning out of the airways is prevented and copious amounts of sputum ensue. Many people with CF bronchiectasis do not bleed and that's a blessing. Unfortunately, I'm not one of them.

By the time of my late CF diagnosis, at 33 years old, I was already having minor episodes of hemoptysis. Of course, at the time, these "minor" episodes were a very big deal. Each streak or clot worried me. Everything is relative – now that I really know of what my lungs are capable. A year later, I had what I consider my first serious bout – feeling like the girl in "The Exorcist", with a stream of blood pouring out of my mouth. That was when I realized the detrimental effects of alcohol and consequent dehydration on my lungs. I now have compiled a list over the years to help me tease out what may be the source of the bleeding.

Everyone is different and you probably have your own list of triggers. My hope is that I can help expedite your discovery of the cause, help you avoid triggers and prevent future episodes of hemoptysis. This is my list: 1. Infection 2. Lack of Rest 3. Lack of Sleep 4. Trauma 5. Dehydration 6. Hormonal 7. Allergic and 8. Stress.

JOHN AND JEANIE HANLEY.

Top on my list always is infection – have I had a recent cold or sinus infection? Has it been a long time since my last tune-up? Have I had more secretions than normal? Even if the answer is, "Yes," I still look over the rest of my list to ponder possible exacerbating factors.

Next to consider is how much rest have I been getting lately. Have I been particularly fatigued? Wearing myself thin? Not allowing myself to sit down, breathe in and exhale comfortably? During these times, I've been running around so much I simply have not been breathing properly – holding my breath and breathing shallowly. Even as I write this, I have to take stock of how I'm choosing to breathe and remind myself to take deep breaths and exhale completely. Even when the activity is very enjoyable as in this moment, the poor cycle of breathing can occur. Checking my breathing helps me find immediate mental respite, if I am feeling fatigued.

When fatigue has set in and the first signs of blood appear, then this is a flashing red neon sign that true rest is now imperative. And I mean resting - as in stop what you are doing that second - lie down or sit or whatever is your most comfortable position, close your eyes and relax. I don't mean, when I get home, or tonight or in a little while. If the bleeding has just begun, I have found this will put an end to it very quickly.

If I've been feeling weak, it's often due to lack of sleep, which is a pervasive issue for many of us. In my case the insufficient sleep is usually due to medications, coughing at night and rapid, shallow breathing. For others it may include sleep apnea, diabetes or any multitude of causes (poor thyroid function, depression, headaches, too much caffeine, smoking, etc). Whatever

Continued on page 29

WELLNESS
NOTE TO SELF

By Julie Desch, MD

I must admit that when I read the theme for this *CF Roundtable*, "If I knew then what I know now", I was overwhelmed with ideas for an article. My brain was spewing thoughts faster than I could process them. This either means that I have learned a lot over the last 50 years, or that I was incredibly stupid as a younger Julie. Or both.

I don't know about you, but I don't like to be told what to do. Ask anyone. So, it makes me a bit nervous to write anything that even hints of preaching to others about how they should handle living with CF. That is the last thing I want to do. Instead, I've decided to simply tell myself what *I* could have done better. Maybe this will be helpful to you young-uns, maybe not. But it's fun to write.

BUB

(This was my family nickname as a kid…actually, they still use it)

Tell Mr. Luther that you don't have a tapeworm...you have CF. He's a 7th grade science teacher...he can handle it.

The next time your friend tells you your fingernails look ugly, tell her there is a direct correlation between the roundness of fingernails and the size of the brain. It's a scientific fact.

Take your enzymes, dummy. Do you enjoy that daily stomachache? (Google whether stomachache is one word or two...it's a fascinating discussion).

When your parents are driving you crazy because they are "overprotective", they are just about always right and you are wrong. This becomes clear only when you are a parent.

Your friends are the best things that ever happened to you at this age. Be grateful they pulled you away from your books (even if they teased you about your nails).

When you feel like hiding and covering your ears when you hear your sister and brother have their CF coughing spasms, don't feel so guilty. It makes total sense.

WOODSTOCK

(This was my nickname on the high school basketball team…at least the one I can disclose here)

Your biggest goal should not be attainment of size Zero. This will

JULIE DESCH, MD

come soon enough, and then you will be mad when *nothing* is small enough to fit your non-existent hips, and you have to shop in the boys department for jeans.

You get only one body...don't trash it.

Nobody really cares if you get a B.

Stop worrying about college. You will look back on high school wistfully, wondering how it passed so fast.

Your parents are smarter than you think. You will learn this some day.

JULIE THE COLLEGE AND MEDICAL STUD(ENT)

Jalapeño eating contests are not good for your stomach lining...even if you wash them down with a margarita.

Stop worrying about "the next *all-important* thing" (i.e. medical school, residency, fellowship, job…). It all works out.

Never ever stop pursuing fitness. This is literally going to be your lifeline.

While it may seem unfair and stupid that you have to do so much to take care of yourself, this is, nonetheless, reality. The sooner you stop fighting reality, the happier you will be. Besides, you might as well get used to it.

Be more open about your CF with friends...they can take it and they will

not run away screaming.

JULIE THE SPOUSE, SIBLING, FRIEND, DAUGHTER

Not allowing yourself to fart in front of your significant other is going to lead to a world of pain.

It is better to be sad beyond belief when your siblings and friends with CF die, than to be afraid to know and love them.

Go to your high school reunions! I hear they were a blast.

Transitions are painful, but change is the one constant in your life, and it always turns out okay.

Be yourself, even if that makes others uncomfortable.

Call your mother.

DRCTNGO

(my ridiculous personalized license plate back in the day of working as a pathologist-I'll let you figure it out)

When it's time to choose a career, pick what you would do for free...then figure out how to make money doing it. Oh, and when that opportunity is thrown in your lap, don't blow it this time.

Pride is the enemy. You know what I mean.

Enjoy your job! You had fantastic training and you are good at what you do. You won't do it for long, so have fun! And even when you have to retire, keep in touch with your work friends.

Even though you don't want to retire early - do it, because you will add years to your life by spending the time to take care of yourself. However, this time, talk to a financial planner first.

MOMMY

They will never be as cute as they are *right now*, no matter what their age; so enjoy this very moment, even if it is taxing and you are completely exhausted and feel like sending them back.

Be absolutely paranoid about their little viruses...always.

Don't fall for the "You're big, you're strong, you're Mommy!" bit. He just wants a toy.

Write down those cute moments and take a lot of pictures. Your memory is going to fail you very soon.

JULIE THE JOCK(ETTE)

Mobility exercises are your friend. Start each workout with these, focusing on your thoracic spine. This is your Achilles heel (and probably yours, too, reader).

Set fitness goals that push you a bit but are not overwhelming.

You are capable of much more than you think. Keep challenging yourself. Think P90X, kettlebell training, etc.

Don't compare yourself to others (especially those who are CFTR-able). Your only competitor is yourself.

Run with CF, not from it.

Interval training is where your gains will be made.

You get to take a day off. Don't feel so guilty. Recovery is WAY underrated.

Get used to starting over every time you get sick. Make it a game, instead of an annoyance. You get to choose how you respond each time.

JULIE THE PATIENT

When it becomes clear that you need IV antibiotics, waiting a few days "to see" (to see what?) is never a good plan.

Do the Vest while lying on your side...it works much better.

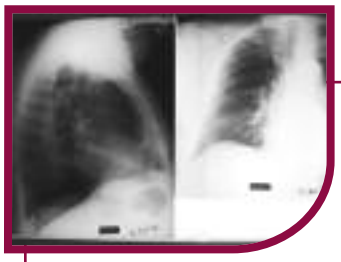
Don't be shy. Tell the floor nurses and hospitalist doctors what you need. You are an anomaly to them (probably not just to them).

When your new (to you) pulmonologist looks at you and says, "Wow, you should be dead!" run, don't walk, to find a different doctor.

Numbers are simply numbers. Median age of survival, FEV_1, TLC, BMI, how much weight you can dead lift, you name it... They don't tell the whole story. What matters is how you feel, what you can do to feel better, and that you are still here to see another day.

So that's it. Just a few random thoughts. What would *you* change if you could do it all over? How can you start making some changes today, so that ten years from now, you won't look back at the last decade with a single regret? I'm going to write Mr. Luther a letter right now. ▲

Julie is 50 and is a physician who has CF. You may contact her at: jdesch@usacfa.org



A DEEP BREATH IN

Now I Know

By Debbie Ajini

I think anyone, with CF or without, could write a long article on what they wish they had known way back when. "Don't get that perm." "He wasn't worth your time." "Buy Apple stock," and so on. I know all those things now! I also know many things about CF and how it has affected my life that I couldn't have imagined when I was younger. The only way to learn these lessons, though, is to live through them and learn from them. Each person's lessons will be different as will our paths in life. Here is what I would tell myself, at various ages, if I could go back in time.

> *"Treating your body with respect, despite it not always doing what you want, will pay off in the long run."*

To my 5-year-old self:

Debbie, it is not your fate to die at 10 like your brother did. CF is an evil disease that prides itself on being completely different for each person. Do not assume your pattern will be the same.

To my 12-year-old self:

When those boys in school are picking on you and calling you names, just know that down the road there is an amazing man waiting to be your husband. A man who will always put your needs before his. A man who will carry you through your darkest hours, CF-related and otherwise. So, just smile and let their insults roll off your back. There are ALWAYS people willing to love those of us with CF, not because we have CF, but because of who we are, in spite of it.

To my 15-year-old self:

Try to be a little more understanding of the Burkholderia cepacia controversy in the early 1980s. The separation of camps wasn't a personal attack on you and those with the bacteria and it probably saved lives. Be thankful they were looking out for you and your friends. Realize there can be a balance between connecting with friends in person and keeping you both safe.

DEBBIE AJINI

To my 17-year-old self I would say:

Get the braces! You don't want to have them as an adult, get them while your insurance will cover part of them! You WILL graduate high school and you WILL get a college degree. You will want straight teeth. STOP assuming you won't live long enough for it to pay off. Think more about finding your passion in life so that you don't realize at 35 you never gave it much thought.

To my 20-year-old self:

Sit with K. and tell her you love her. Hold her while she is scared. Her path took her in a different direction and while it is scary for you to think about, it is more scary for her. Just be with her. You will never regret it, I promise you.

To my 10-, 15-, 20-, and 25-year-old self:

Get moving! I know, I know you have skinny chicken legs and you may not have much grace but moving your body is going to make you healthier in the long run. It will empower you in ways you can't imagine. Treating your body with respect, despite it not always doing what you want, will pay off in the long run. Do right by your body and get it moving, bending and stretching.

To my young-adult self:

Talk more about your disease to the public and to your family. Education is key. I know it is scary to

TILLMAN *continued from page 22*

get up and talk in front of a room full of people, but your story is a good one. There are lessons to learn and things to be inspired by. Help make the change happen.

To my 35-year-old self:

I know this is the worst it's ever been and you and Louie are more afraid than you want to let on. You are going to be okay, you are going to get through this dramatic change in your health. You are going to have a stronger marriage because of it. You are going to learn lessons that will help you become a better wife, friend, daughter, sister and niece. Just take one moment at a time and breathe.

To myself, every year:

Debbie, you are going to celebrate your 41st birthday and more! Stop waiting for CF to run its "typical" course. With CF there is no typical! You have a long life ahead of you. You can fill it with amazing experiences.

To my future self:

Can you believe you have made it this far? You have done such amazing things since you turned 41! That was so long ago. The changes in your health have been so amazing and you are thankful for every day. Each breath is truly a gift. You are checking things off of your "LIST" and adding new items all the time. You live everyday knowing it truly is a fit. Keep doing what you're doing! ▲

Debbie is 41 and has CF. She is a Director of USACFA. Her contact information is on page 2.

David J. Pasta MS, Stefanie J. Millar MS, Joan R. Jacobs MA, Ashley Yegin MD, Wayne J. Morgan MD. Pediatric Pulmonology. Volume 46, Issue 6, pages 545–553, June 2011

The use of dornase alfa for a 2–year period is associated with a reduction in the rate of FEV_1 decline.
http://tinyurl.com/6zl4ep4

Restoration of Chloride Efflux by Azithromycin in Airway Epithelial Cells of Cystic Fibrosis Patients. Vinciane Saint-Criq, Carine Rebeyrol, Manon Ruffin, Telma Roque, Loïc Guillot, Jacky Jacquot, Annick Clement, and Olivier Tabard. **Antimicrobial Agents and Chemotherapy.** April 2011, p. 1792-1793, Vol. 55, No. 4

The aim of this study was to investigate, in human airway cells, the mechanism by which AZM (Azithromycin) has beneficial effects in CF (cystic fibrosis). The authors demonstrated that AZM did not have any anti–inflammatory effect on CF airway cells but restored Cl– efflux.
http://tinyurl.com/3e4h9kp

Pharmacokinetics and Safety of MP-376 (Levofloxacin Inhalation Solution) in Cystic Fibrosis Subjects. David E. Geller, Patrick A. Flume, David C. Griffith, Elizabeth Morgan, Dan White, Jeffery S. Loutit, and Michael N. Dudley. Antimicrobial Agents and Chemotherapy. June 2011, p. 2636-2640, Vol. 55, No. 6

The pharmacokinetics and tolerability of nebulized MP-376 (levofloxacin inhalation solution [Aeroquin]) were determined in cystic fibrosis (CF) subjects. Nebulized MP–376 was well tolerated. Nebulized MP–376 produces high sputum and low serum levofloxacin concentrations. The pharmacokinetics, safety, and tolerability were similar for the two formulations (180 mg. and 240 mg.) received by the subjects.
http://tinyurl.com/3o7oa2w

FYI

Higher risk of hospitalization among females with cystic fibrosis. Anne Stephenson, Janet Hux, Elizabeth Tullis, Peter C. Austin, Mary Corey, Joel Ray. Journal of Cystic Fibrosis. Volume 10, Issue 2, Pages 93-99

Females affected by CF are at a higher risk of respiratory-related hospitalization, which may extend beyond classic clinical measures of disease severity.
http://tinyurl.com/475teqn

The impact of nocturnal oxygen desaturation on quality of life in cystic fibrosis. Alan C. Young, John W. Wilson, Tom C. Kotsimbos, Matthew T. Naughton. Journal of Cystic Fibrosis. Volume 10, Issue 2, Pages 100-106

Nocturnal oxyhaemoglobin desaturation is common in cystic fibrosis (CF) but the effect on quality of life (QoL) remains unknown. Nocturnal oxyhaemoglobin desaturation is associated with impaired QoL, independent of the effects of lung function and awake gas exchange, in stable CF outpatients with moderate to severe lung disease.
http://tinyurl.com/4r534p8

The effect of exercise on large artery haemodynamics in cystic fibrosis. James H. Hull, Les Ansley, Charlotte E. Bolton, James E. Sharman, Ronald K. Knight, John R. Cockcroft, Dennis J. Shale, Rachel Garrod. Journal of Cystic Fibrosis. Volume 10, Issue 2, Pages 121-127

Adult patients with cystic fibrosis (CF) have resting abnormal large artery haemodynamics. Adults with CF have an abnormal haemodynamic response to exercise. This finding has deleterious implications for myocardial performance.
http://tinyurl.com/4c9qrvv

Changing epidemiology and clinical

Continued on page 36

TRANSPLANT TALK

Sun and Skin Do Not Mix

By Colleen Adamson

When I was younger, I did not know nearly as much about sun damage as I do now. I should have been wearing hats and loads of sunscreen when I was outside. I swam and played tennis quite a bit, and I rarely had a hat on. It just wasn't that common or concerning back then. My parents would remind me from time to time, but it was not something people focused on like they do now. My parents were too busy making sure I stayed relatively healthy as a person with CF which, of course, is where their focus should have been.

As a transplant patient, I take a lot of medicines that suppress my immune system. I also incurred (unbeknownst to me at the time) a lot of skin damage when I was younger. Suppressed immune systems and skin damage are not a good combination, because a suppressed immune system enables skin damage to turn into cancer. I've had a lot of experience with this, since my lung transplant in 1998, because in 2004 the skin cancers started to pop up. I have had cancer on my nose, my eyebrow, and my lip. I've had many pre-cancerous spots burned off my neck and hands. My dermatologist calls them classic "Transplant Hands" because this happens to so many people who have had transplants.

I thought that after my lung transplant I would see the transplant clinic doctors more than any other doctors. As it turns out, I see my dermatologist more than I see the transplant clinic doctors! I'm in the dermatologist's office every couple of months, getting spots burned off my hands and neck. Not pleasant – but not terrible. I am currently walking around with a big bandage on the top of my head from recent cancer surgery, and to be honest, it looks weird. I will have a bald spot where the surgery was done, and I haven't quite figured out how to cover it with some kind of comb-over, but I'm working on it. However, you get to a point in life when you realize that how you look after surgery is not all that important. What is important is that the cancer is gone.

COLLEEN ADAMSON UNDER COVER.

It's an interesting dynamic though. I get skin cancer because of the immunosuppressants I take for my transplants. However, I would not even be alive today to experience having skin cancer, if it were not for my transplants. It's all how you look at it; I'd much rather be here and deal with skin cancers than not be here at all.

I wish I had known back when I was a child/young adult that I should have worn hats and sunscreen when I was outside. I don't dwell on it, though. I hope people reading this will realize that skin cancer is extremely serious, and not to be taken lightly. So, please, think of me when you go outside, and don't forget those hats and sunscreens! ▲

> *"As it turns out, I see my dermatologist more than I see the transplant clinic doctors!"*

Colleen is 42 and has CF. She is a Director of USACFA and is the Treasurer. Her contact information is on page 2.

the reason, we do what we can to change any harmful influences and find time to catch some extra zzzzzz's.

The medication that is the major hyposomnia [reduced time of sleep] culprit for me is good old prednisone. Although it is a lifesaver when needed, one of the many side effects is frequent waking at night to cough, going to the bathroom or just what I call "zoning" (as in predni-zone) where my mind is fully alert at 3 in the morning. If my voice didn't carry so well at that wee hour and probably wake up everyone in the house, I'd probably call my niece in Japan to chat and use up some of that energy. Instead I let my family members sleep, try yoga stretches (or imagine myself doing them if I'm too tired to get up), read a preferably mundane article or try visual relaxation by imagining myself sleeping soundly and fully relaxed.

Next issue to ponder is trauma, the kind that causes undue physical strain on your body – either knowingly as in a strenuous workout or rock climbing or, unknowingly, like lifting heavy objects. This has been a big problem, although on the surface, you may mindlessly ask why when it seems clear to just don't do these activities if they make you bleed. For me, if I see a heavy item that needs to be moved, I don't hesitate to lift and move it. Or if there's someone who needs help with unpacking, moving a TV or clearing out a heavy box, my first inclination is to help. Only recently have I made a very diligent effort to remember NOT to be helpful in these situations.

Prior to the bronchial artery embolization procedure, I had no qualms about doing my share or contributing. As a consequence, even after lifting just a few heavy boxes, presumably at the wrong time, I would go home to discover blood in the sputum. I've even had to be a little less gung-ho during upper body workouts, being careful not to mimic climbing exercises. Because a strong upper body is an indicator of good lung health, I have to make sure that I balance pushing myself during workouts and not overdoing it.

Dehydration is another culprit that causes hemoptysis – either from not enough non-caffeinated beverages – or too much caffeine, alcohol, heat, etc. Although I'm very careful about maintaining my hydration well, there are times when I feel that either the weather or a medication is preventing the copious amounts of water that I drink from reaching my lungs and loosening my airway secretions. Increasing the HyperSal 7% as high as two vials three times daily has provided tremendous benefit in augmenting the hydration in my lungs.

> "If I've been feeling weak, it's often due to lack of sleep, which is a pervasive issue for many of us."

Reading a recent *CF Roundtable* Website post about coconut water and rehydration, I decided to try it and a few hours later, lo and behold, I could feel the moisture returning to my lungs. And it has a decent taste! On a scientific tidbit note, Pulmozyme may work more efficiently with magnesium supplementation in those who do not have a good response after inhalation. Coconut water is not quite chock-full but has a great blend of key minerals such as magnesium, potassium and sodium. Coconut water even is used short-term intravenously to hydrate critically ill patients in remote regions of the world where usual sources of hydration are not available.

Other causes of hemoptysis left to consider are hormones, allergy and stress. Premenstrual symptoms are often linked to hemoptysis. I took birth control pills at one point to counteract this effect. Allergic bronchopulmonary aspergillosis (ABPA), an allergy to a fungus that is everywhere in various concentrations, also is a well known cause of bronchiectasis and hemoptysis, affecting 10% of those of us with CF. As the mainstay therapy for this is steroids, the frequent bursts and often chronic use of prednisone is necessary along with all the side effects that follow.

Stress also has to be a serious consideration that requires a bit more reflection, since the more stress we have, the more likely we are to be affected by the other triggers. We all have stress in our lives – our goal is to learn how to deal. Easier said than done! But preparing ourselves for upcoming stressful events or realizing one or some that have recently occurred is the first step in learning how to reduce stress in the future. For me, yoga and meditation have helped immensely in this regard.

I have three rules of thumb when any volume of blood appears with coughing – First I rest; Second, I stay resting; and Third, and most important, I rest some more! Being in tune with triggers of hemoptysis will help prevent future episodes and any kind of rest and relaxation will help before, during and after bouts of bleeding. My experience is that with rest, all the other therapies to treat the hemoptysis will be much more effective and work more quickly. ▲

Jeanie is 49 and is a physician who has CF. She is a Director of USACFA. Her contact information is on page 2.

TRANSPLANT TALK

Sunny with a Chance of Cancer

By Andrea Eisenman

When I look back at how I spent my summers, I cringe. What gives me this uneasy feeling is the fact that I was running around at the beach, playing in the sand, swimming, gardening, doing sports—all without sunscreen. It probably was not until I was 14 or 16 that I heeded my dermatologist's warnings and started using an appropriate SPF. It may have been later. I wanted a tan like all my other friends. Of course, hindsight is 20/20 and at that time, no one knew the damage that the sun could wreak 30 - 40 years later. My parents didn't use sunscreen. If my mom used anything, it was baby oil!

I was not a good tanner. Meaning, I freckled and usually got pretty red, which meant that I then had to treat the sunburn with Noxema. For those of you born after 1986, it was a thick, white moisturizer. Some people used it as a facial wash. It had a very particular smell—menthol. That was my *eau de cologne* most summer nights. Then a few days later came the peeling of skin, which would fascinate me and my friends for hours, trying to see who could pull off the largest amount of skin in one piece. After burning so much, I did start to use 8 SPF.

In my twenties, I was very good about wearing 15 – 30 SPF. But as they say, "Damage done." My mother's family has similar skin, freckly and fair, so a dermatologist was a doctor I saw regularly apart from my CF doctor. I should also mention that I had an abnormal amount of moles on my back. I was told by many doctors that my mole-constellations should be watched as it could cause problems in the future.

CAREN FRIEDMAN AND ANDREA EISENMAN TAKE A BREAK BEFORE FINISHING A 35-MILE BIKE RIDE ON THE NORTH FORK IN LONG ISLAND TO PROMOTE ORGAN DONOR AWARENESS.

Once my family's skin doctor died at age 100, I stopped seeing a dermatologist on a regular basis. It was only after many years, post-transplant, that I was forced to go to a skin doctor for an abscess on my leg. Well, it was not going to be that simple. This dermatologist, Dr. Miller, took one look at me and my moles and had a field day with me

After several biopsies, removal of a big squamous cell and several basal cell carcinomas, I had a date with her every four months. Unless, of course, I saw anything suspicious. Then I should come right away. All this, literally, bit by bit, was hard to deal with. I dreaded my appointments. I just felt I didn't have that much skin left to take. In theory, it was not such a big deal. I have been through worse - major surgery several times. But what bothered me was that each time I had something removed, I had to have stitches. Stitches meant no exercise for about two weeks, while the stitches were in. And it was painful for several days afterward. Even though I received novocaine shots for the pain, I bled tremendously due to the prednisone I took. In total, she removed a few precancerous moles and guided me on the best sunscreen to use. And to <u>always</u> wear it, even if I am just going to the supermarket. Message received.

Dr. Miller and I saw so much of each other, I could tolerate it only because she was amusing and very kind. When she closed her practice, she referred me to an associate who made me get "mapped". (A rather embarrassing procedure to photograph every inch of my nude body for posterity.) That way, if new moles appeared or changed, they could look at my photos and decide whether to biopsy. Following my switch to a new skin doctor and the mapping, I felt a few lumps that scabbed on my head. I would scratch off the scab only to have it reappear, numerous times. At one point, the new dermatologist, Dr. Celebi, put some liquid nitrogen on it. It still came back. Finally it was biopsied and declared a basal cell carcinoma.

I was told I had to see a Mohs spe-

cialist. Mohs surgery was developed by Dr. Frederic E. Mohs. It is microscopically controlled surgery used in obtaining complete margin control during removal of a skin cancer. That way all the cancer is removed and, generally, no more surgery is needed. My mom had had this procedure on her foot. They keep taking pieces until all margins are clear. But this was on my scalp, right in the front of my head and the area had to be shaved. A large triangle was shaved and they made a cut into a big "T" with about 30-40 stitches. I nearly passed out when they showed me the scar and told me how to care for this monstrous wound. I was also told not to exercise for three weeks. Two weeks while stitches were in and one after stitches so I wouldn't get a keloid scar. Not exercising for so many weeks was going to ruin my blood sugars. Aside from glucose and weight controls, exercise helps me maintain my sanity. I knew I was in for a tough time.

I was so traumatized by this procedure because it hurt so much for at least three days. I had to take Tylenol 3 and still I had throbbing pain. After it healed, I noticed another scabby area on the apex of my scalp. It was biopsied and then I was told I needed another Mohs procedure. I was upset and the doctor could feel my pain. She said, "Take a break and come back in three months."

> *"I am pretty sure that running around all those years without sunscreen, hat or a care in the world led me to all these skin cancers."*

I have since "graduated" from my second Mohs experience. I am pretty sure that running around all those years without sunscreen, hat or a care in the world led me to all these skin cancers. My genes helped too. Plus, on top of it all, I am immuno-suppressed due to my lung transplant. So, I may have had all these cancers but without being immune compromised, they might have become apparent in my 60s, if I lived that long, instead of my 40s.

The reason I share my experience is to urge everyone to be aware of the risk of sun exposure as well as the importance of wearing sunscreen, sunglasses and possibly a hat - especially if you have fair skin and light eyes, freckle, or are on immuno-suppressants. It is important to see a dermatologist early, before you have symptoms. And don't forget to have your scalp checked. They can take a look and assess your risks of skin cancer and can get a "baseline" of your skin before you have problems. Catching things early before they develop into dangerous cancers, such as melanoma, is key.

If only I had known then, what I know now, I might have most of my original skin. ▲

Andrea is 46 and has CF. She is a Director of USACFA and the Executive Editor/Webmaster. Her contact information is on page 2.

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Hiking The Grand Canyon

By Cynthia Dunafon

In May, under a gibbous moon, I walked to the rim of the Grand Canyon and looked out. The entire canyon was illuminated in white light. The Battleship, a ridge overlooking the Bright Angel Canyon and familiar to most park goers, was clearly visible. Several great temples of stone on the north side of the Colorado River cast their shadows: Isis, Cheops, Buddha and Brahma. The white Coconino sandstone was aglow, forming a horizontal band of light just below the north rim, ten miles distant. Not many stars could compete with this splendor, yet Draco arched across the southern sky to remind me that I was in the abode of dragons.

CYNTHIA DUNAFON AT PLATEAU POINT ON THE BRIGHT ANGEL TRAIL, GRAND CANYON.

Ramp Up

Three years ago, I spent an afternoon at the Grand Canyon with my sister. We sat and watched hikers emerge from below the rim – tired, dusty and elated. I promised myself that I would return, soon, and hike those trails. Finally, this past May, I spent two weeks there. It was an exploratory trip, dedicated to day hiking and figuring out how this middle-aged person with CF, in good-but-not-great condition, could hike successfully in the canyon.

Since I have called the Midwest home for many years, I knew that I would need a few days to adjust to the elevation, which is over 7000 ft at the south rim. What better way to justify a stopover in nearby Flagstaff than to visit Mars Hill, the site of the famous Lowell Observatory? Getting there from my hotel involves a short walk (a mile or so) and a 300 ft climb. All too soon, I'm gasping for air and need to rest a few times before reaching the gate. I wonder if I'll be up to any hiking, but then I've been here for only about 24 hours.

Onward and upward.

I find myself getting thirsty frequently as I walk around the Observatory. No wonder, last week Flagstaff recorded its lowest dew point ever: -0.6!

Percival Lowell, the founder, spent much of his scientific career studying the solar system and mapping the surface of Mars. Although the later Mariner missions disproved his theory of canals (and an intelligent race of canal builders), his dedication to Mars mapping caught the public imagination. Those of us who follow the explorations of the Mars Rovers, or who simply enjoy a good yarn about invasions from the Red Planet, have Lowell to thank for his legacy. After the various historical tours available during the day, a cloudless night sky ensures a clear view of Saturn and her moon Titan through Lowell's century-old, 24-inch, refracting telescope.

It's a Tobramycin month and my bronchospasms are increasing. After a bad night of coughing, I forgo the morning dose. A long shuttle ride to the Grand Canyon awaits and I don't feel like ruining the ride for the other passengers with my drug-induced side effects. Sorry, doc. I do the rest of my aerosols plus 40 minutes of Vest before heading to the shuttle stop.

To continue the acclimation process after arriving at the Grand Canyon Village, I spend the next two days walking the rim trails, which are splendid in their own right. I love how the colors in the canyon change over the course of the day, and I get a taste of the unpredictable weather with a late afternoon hailstorm on

the day after my arrival. These preliminary hikes allow me to test out my clothing, pack weight and water use. By the time I take my first step below the rim, I have spent a total of four days acclimating to the elevation, walking almost 35 miles. Onward and downward.

South Kaibob Trail

The South Kaibob Trail is steep, scary and spectacular – just what one would expect of a Grand Canyon trail. I keep my eyes on the trail during the first set of switchbacks down an almost vertical cliff face to keep me from thinking too much about the empty space nearby. When I feel more confidence in myself and in the width of the trail, I begin to take stock of my surroundings. After a mile or so, the trail follows a ridge that juts out into the main canyon. It's breathtaking. There are sweeping vistas on either side competing for attention. You can't take it all in. Your eyes search for a particular rock, shadow or bird to provide a sense of scale.

The first rest stop, Cedar Ridge, is about 1000 ft down and 1.5 miles in. Here, the trail widens out into a broad expanse of deep red shale with scattered juniper trees and other scrub plants. An aggressive squirrel heads straight for the opening of my backpack and has to be removed bodily. I make a mental note for my hiking journal: get better food containers. So far, I'm feeling good. With the help of hiking poles, I've descended the steepest parts of the trail very slowly to keep from jarring my knees too much. A scrub jay surveys the canyon from a boulder nearby.

I soon push on to my final destination, Skeleton Point, which is another 1000 ft down and 1.5 miles farther along the trail. That's not a long hike on level ground, but I don't know how long the ascent will take or how hard it will be. The goal is not to exhaust myself, but to find out what a 6-mile hike with a 2000 ft climb feels like. If I want to do more, I'll plan a longer hike tomorrow. The park service frequently reminds visitors that "what goes down must come up". There are no mules-for-hire waiting in the lower reaches of the canyon to take you back up to the top. Know your strength; know your pace. That's what I'm here to assess.

Skeleton Point is a good-sized promontory on the north end of O'Neill Butte. It's scattered with tall, majestic Utah agave, hedgehog cacti and small, bright wildflowers. From this vantage point you get a good sense of the immense Tonto Platform, a sloping terrace of gray and green limestone that separates the upper layers of the canyon from the Precambrian crags of the inner canyon. The meandering Tonto Trail is visible in places. Wotan's Throne and Vishnu dominate the western horizon. A hiker from Holland strikes up a conversation, and we look at my topographical map for the names of other temples and side canyons.

The hike back to Cedar Ridge goes well. This section of the trail contains a series of straight, sloping trails with just a few switchbacks towards the end – not nearly as steep as the first section of the trail. It's warm in the sun and I discover how small changes in light, temperature and the slope of the trail impacts how I feel moment by moment. A woman from Maryland notices my Chicago Respiratory Association T-shirt that I got climbing the stairs in the Hancock Tower last February. How even and predictable those flights of stairs seem to me now.

I take the final switchbacks very slowly. Walking at this pace has allowed me to maintain an even rate of breathing so that I make it back to the top without hyperventilating, an important discovery. At the right pace, my musculo-skeletal strength plays a greater role in my hiking performance than my lung capacity. Like many people with CF, I assumed my lungs would be the weakest link, but "it ain't necessarily so!"

A longer note from my journal: In the high desert, the sweat evaporates right off your skin. You might think that you're not even breaking a sweat, but you are. So, it's hard to gauge how much water you're losing as you hike. I also lose moisture by breathing through my mouth. A higher breathing rate than normal exacerbates this problem. At the park, all hikers are not only encouraged to drink lots of water, but also warned not to neglect electrolyte replacement. That's especially true for CFers. For my day hikes, I focused on hydrating my body on the descent and carried a two-liter bottle of water for that purpose. For the ascent, I had a one-liter bottle of water with an electrolyte replacer (I use Vitalyte and a twist of lime for flavor). A few swallows of this at each rest stop was enough to keep me going. In addition, I carried a small zipper bag of sea salt. When the sun was bearing down, I had a pinch or two of salt when I stopped for drinks/snacks. It's less bulky than carrying pretzels, and my body thanked me for it.

Bright Angel

My longest hike on the Bright Angel Trail was to Plateau Point, an amazing lookout point on the Tonto Platform, 3000 ft down and six miles in. Like the South Kaibob, this is a "corridor" trail – meaning that it is accessible from within the park and is maintained (and patrolled) by the park service. Don't expect isolation on this trail; it's the most popular one in the park. The high volume of traffic gives the trail a unique character. The intrepid hiker must carefully

Continued on page 35

CONVERSATION CORNER

CF Can Be Irksome – Just Deal With It

By Walter Bartholomew

I thought I would share my thoughts on a question I often receive regarding having cystic fibrosis, now that I am 38, going on 39. The answer to this question would clearly be different for others who suffer from this same disease. It would depend on numerous factors such as health, both physical and mental. Regardless, I felt my answer may help people better understand how I deal with or accept my life with CF, and how this answer might help others who often don't understand how to help or interact with this disease.

Question: "How do you live with CF and continue to fight everyday?"

This question can simply be answered by many by just saying, "I deal with it." During my childhood CF was perceived by me as more of a chore than a detriment to everyday life - meaning, I didn't have sacrifices or limitations on my activity or stages of ups and downs. My father ingrained in my schedule the routines required to stay healthy, so I never had a bad childhood with my CF. As I got older, and during my college years in the early 2000s, I started to feel the impact on my health. My breathing got much shorter and routines to stay healthy took longer. Now that I am 38, and have been in decline over the past 10 years and fighting to stay "baseline", I answer this same question completely differently.

I thought about and researched what word best describes how I live with CF every day. The one word that is kind of funny and gets my point across is *irksome*. Every day I awake it's a chore to just start moving around. I have to take medication before I do anything. I have to watch what I eat and drink, what activities I partake in, how I travel and many other things people take for granted and would not even think to plan. Given I have two kids, work full time and have a family household, tons of planning and control are required to function each day. It's so easy to lose sight of CF with everything going on around you. I have to plan each day around my CF - from work, to kids, to family functions.

To say it's irksome is just a simple answer, but it's much more than that. On top of it all, we hide how we feel. We sit through meetings, go to kids' sports games, talk on the phone, go out socially - while the whole time we could be experiencing pain breathing or feel like crap. People with CF tend to hide how they feel as it's hard to explain to anyone what is causing the discomfort. It's funny but, CF is an hour-by-hour and even minute-by-minute disease. You could be feeling great one second and the next feel like crap. It fluctuates depending on where you are, what you're eating and drinking, the temperature or just because CF wants to change course. As soon as you think you have things steady and under control - things change. How on earth do you answer this question? It would be different each day, each hour and each minute. That's what is irksome; the lack of control. The more you try to control it, the more it fights back, it seems.

CF can also provide a blessing that no one else on earth can experience - the sense to enjoy every day, because you never know when your time might be close to leaving this great earth. You never quite know when your lungs might quit or heart might stop, but then most people don't. But we with CF are reminded with every breath that this could happen at any time. We block this out most days, but we also try to experience the greatest this life has to offer: our relationships with family and friends and even nature. Movies and songs have a different meaning. The scenes that go on around us are easily missed by the busy lives we lead. Now I find myself watching more of life's moments. It can be as simple as watching kids play sports to someone sitting on a bench in the park. You spend your day searching and searching for something happy and something special, while at the same time you fight your CF, whether you feel like crap or even when you feel just OK.

So when you ask someone who has CF how he fights each day or how he feels, don't be surprised if you get a one-word answer. Because they may not know either, and it will likely change the next hour, anyway. I hope this was helpful in some small way to people with CF and people who interact with someone suffering. ▲

Walter is 38 and has CF. He lives in Easton, PA with his wife and two children.

navigate a trail worn down into uneven ridges and grooves by untold numbers of boots and hooves. But that's only the beginning. There are the quick (rim to rim runners), the slow (yours truly), the young and the old (both of whom hike faster than the rest of us), the tour groups (from all over the world), the confident ("Das ist nicht so steil!"), the misinformed ("It must be safe, there are old people on the trail."), the grizzled, the barefoot (I'm not joking), the photographers who stop suddenly for that perfect shot and, of course, the mules who always have the right of way. Veteran hikers practice a complicated form of social etiquette, but for the most part, I just tried to stay out of the way of faster hikers. On level ground, I have a strong stride and am used to passing up slower people, so it was hard to be the one that everyone had to pass. But, I kept focused on the larger goal of even breathing, balanced use of my leg muscles and reserving enough energy to climb back up without sounding or looking like I needed medical assistance.

The log steps were harder to traverse than I expected. Deep depressions worn into the trail meant that you stepped down with one leg before lifting the other leg up over the log and then down, way down, onto another uneven patch of trail, all before bringing the first leg over to join it. After a few hundred repetitions, you become increasingly adept at finding side paths in the dirt that will allow you to avoid even one or two such steps.

An amazing coincidence occurs on my way down the trail. I hear someone call out my name. It's a student of mine from Chicago who is hiking up from the river.

Halfway between the rim and the river is an oasis called Indian Gardens. There's a campground here, a ranger station, and a lively watering hole for hikers. Some are coming up from the river, some are on their way down, and others, like me, are here for a day hike. Everyone has a story to tell about the canyon. The tall trees provide welcome shade after the long stint on the trail. (Large temperature gauges are posted along with the sign: "This is your brain in the sun!" to remind people of the disorienting effects of heat stroke.)

Plateau Point is a mile and a half from Indian Gardens. This section of the trail is fairly level; it takes you out of Bright Angel canyon and travels across the wide expanse of the Tonto Platform – right up to the edge overlooking the inner canyon. Here, I get my first close up look at another world: miles of basement rock, the ribbons of pink granite running through black schist, and the muddy Colorado River in the midst of it all. I can look almost straight down to the river 2000 ft below – dizzying. Three rafts run a stretch of white water far below (mile 89 for river runners) and I delight in the antics of white-throated swifts not far from my rocky perch.

Reluctantly, I head back. When I arrive back at Indian Gardens, it's past midday and getting hot. Even though I don't want the extra weight in my pack, I decide to fill up my water bottles once again. When I finally get to the rim, I find that I have enough energy to take the shuttle to the grocery store for sandwich ingredients. I'm ravenously hungry and feel like celebrating. The tuna salad/cheddar cheese sandwich and chocolate ice cream bar (consumed on a bench overlooking Bright Angel Canyon) seem like the best meal ever.

Over the course of two weeks, I hiked about 120 miles and climbed a total of 15,000 ft in the canyon. I kept a detailed journal of what I did each day, what CF issues impacted my hiking, and a long list of "lessons learned". Because, of course, I plan to return. Stay tuned. ▲

Cynthia is 47 and has CF. She is a Director of USACFA and is the Vice President. Her contact information is on page 2.

Cartoon by Bonnie Bleiweiss

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.

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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

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TILLMAN *continued from page 27*

issues arising in an ageing cystic fibrosis population. Michael D. Parkins, Vicky M. Parkins, Jackie C. Rendall, Stuart Elborn. Therapeutic Advances in Respiratory Disease. April 2011 vol. 5 no. 2 pp 105-119

Improvements in the quality and implementation of medical care for individuals with cystic fibrosis (CF) have resulted in a dramatic improvement in survival. Many of these strategies have focused on the effective management of pulmonary disease which has delayed its manifestations into later years. With an increasing number of patients surviving to later years, the impact of chronic inflammation and nutritional compromise on other organ systems over a lifetime are increasingly manifest.
http://tinyurl.com/46f275l

Association between genotype and pulmonary phenotype in cystic fibrosis patients with severe mutations. A. Geborek, L. Hjelte. Journal of Cystic Fibrosis. Volume 10, Issue 3, Pages 187-192 (May 2011)

Patients with two class I mutations had lower lung function compared to the group with either a combination of class I and II mutations or two class II mutations. Cystic fibrosis patients carrying two class I mutations risk developing more severe lung disease compared to patients with at least one class II mutation.
http://tinyurl.com/3tpyfsy

GENE THERAPY

Current Status and Future Directions of Gene and Cell Therapy for Cystic Fibrosis. Griesenbach, Uta; Alton, Eric. BioDrugs: 1 April 2011. Volume 25, Issue 2, pp 77-88

Early pre–clinical studies have recently been initiated to address cell therapy–based approaches for cystic fibrosis. In this review, the authors discuss recent developments with viral and non–viral vectors and cell therapy, and provide an update on clinical gene therapy studies.
http://tinyurl.com/3wzweaf

NEWS RELEASE

Phase 3 STRIVE Study Of VX-770 Showed Durable Improvements In Lung Function And Other Measures Of Disease Among People With A Specific Type Of CF

Vertex Pharmaceuticals Incorporated announced the final results from its pivotal Phase 3 STRIVE study that evaluated VX-770, a medicine in development that targets the defective protein that causes cystic fibrosis (CF). STRIVE was designed to evaluate VX-770 among 161 people 12 years or older with a mutation known as G551D in the CF gene. Approximately 4 percent of people with CF have at least one copy of the G551D mutation. Data from the study showed rapid improvements in lung function (FEV_1) that were sustained through 48 weeks among those who received VX-770, compared to those treated with a placebo. Significant improvements in all key secondary endpoints were observed among people who received VX-770. Adverse events that occurred more frequently among those treated with VX-770 compared to placebo were headache, upper respiratory tract infections, nasal congestion, rash, dizziness and bacteria in the sputum.
http://tinyurl.com/5v2stcj

Vertex Plans To Provide Access To Potential CF Therapy VX-770 For Patients With Critical Medical Need

Vertex Pharmaceuticals Inc. announced a plan to provide VX-770, a CF medicine in development, to people with the G551D mutation who are in critical medical need and may benefit from treatment prior to potential approval of the drug from the U.S. Food and Drug Administration (FDA). Pending FDA review and

approval, Vertex expects to open the program at clinical sites in the United States as early as July.
http://tinyurl.com/686rjyr

Drug Candidate From Hen's Eggs Might Replace Antibiotics For Patients With Cystic Fibrosis

Antibodies from hen's eggs (IgY) may shortly come to revolutionize the treatment of patients suffering from cystic fibrosis. Most patients with cystic fibrosis are affected by severe and ultimately chronic infections with Pseudomonas aeruginosa bacteria. To date, the only treatment has been repeated courses of antibiotics, with a high risk of side effects and bacterial resistance. The IgY-method, using antibodies from hen's eggs, prevents infections and is also interesting as an alternative treatment to antibiotics in the struggle against other bacteria where the trend is an increase in the resistance. The drug candidate, Anti-Pseudomonas IgY, is a solution with antibodies specifically targeting Pseudomonas aeruginosa. The hen is vaccinated with the bacteria and will thereby start producing antibodies that are transferred to the egg in a high concentration. When the patient gargles with the solution, the antibodies will attack the flagella of the bacteria preventing them from attaching to the cell walls in the patient's mouth and throat. This in turn prevents the bacteria from further infecting the lungs. Anti-Pseudomonas IgY is a water based solution, free from any other additives. As a natural and safe solution it is especially suited for long-term, prophylactic treatment.
http://tinyurl.com/6hhkaf9

Genes Tied to Severity of Cystic Fibrosis Identified

The severity of cystic fibrosis, a life-threatening hereditary condition that affects the lungs and digestive system, seems to be influenced by genetic variations. The researchers were able to identify a region encompassed by two genes on chromosome 11 linked to severe cases of the disease. A second region on chromosome 20 was also identified. Continued study of this region revealed five genes that are turned on in respiratory cells, some of which are known to cause inflammation. Identifying the genes that influence the course of disease may enable prediction of disease severity and, most importantly, the customization of treatments for patients with unfavorable genetic modifiers.
http://tinyurl.com/3d2mu97

Unexpected Help From Cystic Fibrosis Bacteria - Antibiotic Resistance

A bacteria which infects people with cystic fibrosis could help combat other antibiotic-resistant microbes. Researchers have discovered that antibiotics from Burkholderia are effective against MRSA and even other cystic fibrosis infecting bacte-

Continued on page 38

Calling All Writers

Have you written an article or story for *CF Roundtable?* If not, why haven't you written? Are you concerned that you may not be a great writer? Don't let that stop you. We have people who will work with you, on your article, to make it the best it can be.

Are you concerned because you can't think of a topic? How about if we give you a few ideas to start with? Here are some titles that go from head to toe and might pique your interest to write. Remember, these are only suggestions. You may come up with entirely different ideas and that is fine with us. All we ask is that you write about your experience with CF.

Here are a few possible topics for your use: headaches; understanding what you hear; pain(s) in the neck; arm twisting; the case at hand; a breath of fresh air; gut reaction(s); pain in the butt; oh, my aching back; getting hip to a subject; standing on one's own two legs; at the foot of the problem; toeing the line; my sole responsibility. As you can see, these are humorous suggestions that are meant to give you some ideas. You need not use any of these, but you may, if you wish. For other ideas, check out the Looking Ahead section on page 3. All submission dates for the coming year are posted there.

We ask that all submissions be typewritten. If you want to e-mail your submission, please have it in Microsoft Word or a similar program. You may send your submissions to: cfroundtable@usacfa.com
or to
USACFA
PO Box 1618
Gresham, OR 97030-0519.

TILLMAN *continued from page 37*

ria. Around one quarter of Burkholderia bacteria have very strong antibiotic activity on multidrug-resistant pathogens such as MRSA. One particular strain, Burkholderia ambifaria, was found to produce two very potent antibiotics active on resistant bacteria, in particular *Acinetobacter baumanii*.
http://tinyurl.com/65yq46e

Insmed's ARIKACE® Demonstrates Sustained Benefit Through Six Cycles Of Treatment For Cystic Fibrosis Patients With Pseudomonas Lung Infections

Insmed Incorporated announced positive data through six treatment cycles (72 weeks total duration) of its Phase 2 clinical trial program for ARIKACE® (liposomal amikacin for inhalation) in cystic fibrosis (CF) patients with Pseudomonas lung infections. The open label Phase 2 study was designed to evaluate ARIKACE over multiple treatment cycles in CF patients with Pseudomonas lung infections. The study enrolled 49 patients to receive ARIKACE 560 mg once daily for 28 days of therapy, followed by a 56-day off-treatment observation period. ARIKACE was administered using an optimized, investigational eFlow® Nebulizer System. The data demonstrated that ARIKACE, delivered once-daily for 28 consecutive days, followed by 56 days off-treatment, for a total of six cycles, resulted in statistically significant improvement in lung function that was sustained over a 72 week period.
http://tinyurl.com/3llmgbx

Phase 2 Study Of Two Potential CF Therapies - VX-770 And VX-809 - Shows Promising Results In Patients With Most Common Mutation

Vertex Pharmaceuticals Incorporated and the Cystic Fibrosis Foundation announced promising results from an ongoing Phase 2 study evaluating combinations of VX-770 and VX-809, potential medicines designed to treat the defective protein that causes cystic fibrosis. The study enrolled 62 people with two copies of the most common CF mutation, known as Delta F508. The trial lasted three weeks. Participants took VX-809 for two weeks, and VX-809 and VX-770 together for a third week. Patients who took the drug regimen showed a positive change in sweat chloride levels. The findings suggest that VX-809 and VX-770 together improve function of the defective CF protein, known as CFTR.
http://tinyurl.com/5wmf2ku ▲

Laura is 63 and has CF. She is a Director of USACFA and the President. Her contact information is on page 2.

Call to All Artists

If you wish to submit art that expresses your feelings about CF or anything on your mind, please send photographs of any media: paintings, illustrations, collages, drawings, sculpture, etc. to:

cfroundtable@usacfa.org. or you may mail them to:

USACFA
PO Box 1618
Gresham, OR 97030-0519.
Please include your name and contact information.

Webcasts Sponsored by the CFF

Check in regularly at www.cff.org for information about Virtual CF Education Day Webcasts, sponsored by the CF Foundation. The January Webcast will feature experts discussing cystic fibrosis fertility and pregnancy issues. Check out the Website for more information: www.cff.org.

CF Living

CF Living

Check out the new *CF Living!* As many of you may know, *CF Living* is an educational program that offers information and support for those living with or caring for someone with cystic fibrosis. Genentech is proud to offer you this updated form of support, and we encourage you to check out all the new features on the site that are now available. The new *CF Living* is designed to help you work more closely with your Care Team, learn about treatment options, and provide interactive educational resources so you stay informed. Enroll today at: https://www.cfliving.com/ to begin taking part in this informative program!

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I am interested in CF Roundtable because I am a: CF ADULT _____ BIRTHDATE _____

OR I am a: Parent ____ Relative ____ Friend ____ Medical/Research Professional ____ Caregiver ____

A yearly donation of $15 for individuals, $25 for non-US addresses (US funds only) and $25 for institutions is recommended.
CF Roundtable is available free of charge to those who are unable to donate at this time.

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Everyone must complete a subscription form annually to be included on our mailing list. Please check mailing label for renewal date.

HOW TO KEEP YOUR SUBSCRIPTION UP TO DATE

Do you wonder when your *CF Roundtable* subscription is due for renewal? Have you wondered how to tell if it is time to renew? Look at your mailing label. Immediately after your name, there should be a date. That is your renewal date. (On the example, you can see that Kathy is due to renew her subscription in May 2012.) If there is no date or it says (TIME TO RENEW), your subscription is due for renewal.

KATHY RUSSELL 5/12
4646 NE DIVISION STREET
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If it is time for you to renew your subscription, you should have received a reminder with your newsletter. Please be sure to complete and return that form to us. You can help to keep our postage costs down by letting us know your current mailing address. To keep our records up to date, send us a notice when you move. (Any issue of the newsletter that is returned to us costs us about two-and-a-half times the first-class postage rate for that piece. Currently that runs about $3.73 per returned copy.)

Thank you for helping us with this.

IMPORTANT CHANGES

You may have noticed that USACFA and *CF Roundtable* have experienced some changes. There are new officers and board members in our organization. *CF Roundtable* has a different column: "Editor's Notes" rather than "A Word from the President." Also, USACFA has expanded to the point where we have two separate addresses for different purposes. We ask that all correspondence that does not include any money in it be sent to the old PO Box number and that all mail containing money of any kind be sent to the new PO Box.

When sending in a subscription or donation of any amount, send it to: **USACFA, PO BOX 151024, ALEXANDRIA, VA 22315-1024.**

All articles, general inquiries, comments, questions, or praise should be sent to: **USACFA, PO BOX 1618, GRESHAM, OR 97030-0519.**

REMINDERS

- Please notify us immediately of any address changes. Returned mail wastes money and delays mailings.
- To keep our mailing list current, we require the completion of a subscription form every year, even if you are unable to make a donation. Your address label indicates when it is time to renew. Subscriptions stop automatically if not renewed regularly.
- 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**.

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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/