CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
SUMMER 2012

Living In Two Worlds

By Jen Eisenmann

I live in two worlds. There's the healthy world and the CF world. I used to try to cling to the healthy world and avoid the CF world out of fear. But now I accept both as a part of my life.

In the healthy world, I go out in public. I can generally blend in while in public. I don't "look" sick and often I can hold back my cough or make my cough sound like a sneeze and will get your basic, "Bless you." I take my pills so fast before I eat that it usually goes undetected. I like the fact that I can blend in and don't get strange looks from people and hate that people who are more obviously disabled are treated differently. Since I was little, I have always tried to offer people who are ill or handicapped the same decent human respect that everyone deserves – I look them in the eyes and smile - they do not need my looks of pity nor my looks of wonder.

In the healthy world, I have family and friends who are painfully aware of the two worlds I live in. They see me in public, where I keep my disease at bay, but in private, they hear my uncontrollable coughs and gasping for air; they are aware of the frequent stomach pain and digestive problems, and they see me get tired easily. Although they cannot relate (you can really "relate" only with someone who has had the same or very similar experiences as you), they are wonderful and understanding and sympathetic. They don't treat me differently, but respect my limitations.

In the healthy world I also have acquaintances. These are the people I work with, volunteer with, attend book club and other monthly meetings with and have random encounters with. This is the hardest group for me to figure out how to relate to in terms of my CF. They know me well enough to know I have CF but are not close enough for me to share my struggles. They worry about me

JEN EISENMANN

from time to time, especially if I am absent for being sick, but they really don't have any idea how sick I am or what I go through, because they "see" me as healthy, energetic Jen. When they find out I only work two after-

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EDITOR'S NOTES

There have been some more changes at USACFA. Three of our Directors, **Maggie Williamson**, **Cynthia Dunafon** and **Kathy Russell**, chose not to run for reelection. Three new people, **Marie Fioccola**, **Jennifer Hale** and **Stephanie Rath**, have been added to the board. You may read all about them on pages 14 & 15. Maggie, Cynthia and I all enjoyed our years as Directors and will stay involved in our CF Community. We wish the three new Directors well and hope that they will have a wonderful tenure on the board.

Another change is that, for the first time, we will be sending renewal notices to our subscribers. We all know how easily renewal times can slip by so we hope that these notices will make remembering easier for all.

This issue has a lot of good information about living with CF. **Beth Sufian** covers important topics including the Affordable Care Act and the U.S. Supreme Court, Medicare, Medicaid and private insurance, in *Ask The Attorney*. In *Spirit Medicine*, **Isabel Stenzel Byrnes** talks about the "not enoughs" of our lives. **Andrea Eisenman** writes, in *Transplant Talk*, of the minutiae of life that can pile up on us. **Julie Desch,** in *Wellness*, tells of her experiences with getting knocked down by CF and coming back from it. As usual, **Laura Tillman** gives us all the latest *Information from the Internet*.

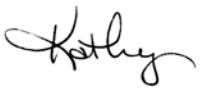
There are three articles in *Voices from the Roundtable*. **Hal Soloff** writes of his 81 years of life and what he has learned. **Annie Opatz** talks of CF and death. On the front page, **Jen Eisenmann** tells us about the two worlds that many of us inhabit.

Paul Feld reviews Andy Lippman's latest book. **Mark Levine** writes about how he spends his time while doing treatments. **Jennifer Hale,** *Coughing With A Smile*, discusses ways to stay in touch when you have CF. *In the Spotlight* features **Jordan Herskowitz** and how he entertains with "Growing Up Jordy Pordy".

Check out the notice about CFTR2, where you can get information about specific CF mutations, on page 9.

As always, we hope that you will look at the future Focus topics, on page 3, and see if there is a topic on which you would like to write. If you are an adult who has CF, we want to hear from you. Give it a try.

Until the next issue, stay healthy and happy,



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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

Lawrence Lafary
Riverton, IL
72 on December 5, 2011

Laura Mentch
Bozeman, MT
59 on May 21, 2012

Debra Radler
Roselle, IL
50 on May 31, 2012

Marion Rojas
Oakland, CA
75 on June 7, 2012

Kathy Russell
Gresham, OR
68 on April 17, 2012

Hal Soloff
Norwich, CT
81 on April 9, 2012

Wedding

Arthur and Brandie Herron
Sacramento, CA
4 years on May 21, 2012

Debra and Adrian Radler
Roselle, IL
5 years on June 8, 2012

Transplant

Greg Briggs, 58
Jacksonville, FL
Bilateral lungs
15 years on April 30, 2012

LOOKING AHEAD

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

Summer (current) 2012: Respiratory Therapy Activities.

Autumn (November) 2012: Organ Transplant - Has It Met My Expectations? (Submissions due September 15, 2012.) Tell us about your life since transplant. What was your quality of life before and after? How has your life changed? Would you consider doing it again? Are there things you would do differently, if you had it to do over?

Winter (February) 2013: Osteoporosis, Osteopenia And Other Skeletal Issues. (Submissions due December 15, 2012.) Tell us of your experiences with your bones. Do you have osteopenia or osteoporosis or any other skeletal problems? How are they being treated? What tips do you have for our readers?

Spring (May) 2013: Gastro-intestinal Issues. (Submissions due March 15, 2013.)

ASK THE ATTORNEY

Answers to Questions Regarding Insurance Coverage

By Beth Sufian, JD

In the past three months, the CF Legal Information Hotline has received over 1,000 calls from people with CF, their family members and members of the CF healthcare team. The questions are often complex and require research of current laws and regulations that affect the ability of people with CF to obtain Social Security benefits, government benefits, modifications to the educational environment and issues related to discrimination in the workplace. Adults with CF continue to face a variety of issues whether at school or in the workplace and often need information on government benefits and access to health insurance coverage. Questions posed by *CF Roundtable* readers are answered below.

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. What will happen if the United States Supreme Court strikes down the Affordable Care Act?

On June 28, 2012, the Supreme Court of the United States rendered its decision on the constitutionality of the Affordable Care Act, also known by many as Health Care Reform. In a 65-page opinion written by Chief Justice Roberts, the Supreme Court upheld the constitutionality of the Affordable Care Act and its provision of an individual mandate, which requires all citizens to obtain health care coverage starting in January 2014. The underlying purpose of the law is to extend health coverage to all Americans, particularly those individuals with pre-existing conditions who are not guaranteed coverage under existing federal law. The law will help people with CF who otherwise have no access to health insurance coverage through an employer, parent or spouse's group policy.

The sections of the Affordable Care Ac that have already gone into effect will continue to be in effect. Thousands of patients with cystic fibrosis have already benefited from the sections of the Affordable Care Act that are already in effect. Specifically the law's elimination of lifetime maximums and the requirement that parents be allowed to keep dependent children on their policies until the child reaches the age of 26. In addition, the law currently allows a child under the age of 19 to purchase an individual policy regardless of the child's health. Under the provisions of the ACA that will go into effect in January 2014, any individual regardless of age with a pre-existing condition will be allowed to purchase an insurance policy. A significant aspect of the law is that pricing will be regulated by a "health insurance exchange", which will not only allow those with pre-existing conditions to purchase a policy, but will offer such policies at competitive prices. For more information about the Affordable Care Act, go to http://www.healthcare.gov/law/index.html or contact the CF Legal Information Hotline.

BETH SUFIAN

2. I am eligible for Medicaid but currently I am eligible for health insurance coverage under my mother's insurance policy offered by her employer. If I decline Medicaid coverage can I apply for Medicaid in the future?

A person may apply for Medicaid if he/she meets the Medicaid eligibility criteria in his/her state. A person must have low income and low assets to be eligible for Medicaid. The fact that a person does not have access to health insurance does not automatically make someone eligible for Medicaid There are only a few states that allow people over the age of 18 (in some states age 19 and in some states age 21) to obtain Medicaid if the person is not also receiving SSI benefits. Before moving to another state, an adult who receives Medicaid benefits and is not on SSI

should make sure that the other state offers Medicaid to adults who are not on SSI.

3. What could happen to a person who gives false information about income to Medicaid and the Social Security Administration?

When a person gives false information to Medicaid or to the Social Security Administration the person is providing false information to the federal government. There are criminal penalties for providing false information on applications for federal benefits or on requests for information by a federal agency. State Medicaid agencies usually find out when a person has provided false information. If the state determines that false information was provided and therefore the person was not actually eligible for Medicaid benefits, the Medicaid agency will ask all medical providers to reimburse to the state any funds paid for the medical care of the person. Medicaid will then request repayment from the person who provided false information for any money paid by Medicaid for medication or medical care.

4. If I decline Medicare Part B can I enroll in Part B at any time in the future?

Medicare Part B covers outpatient services such as physician services, lab work, pulmonary function tests, sputum cultures, durable medical equipment (such as a nebulizer or machine), some inhaled medications and other services. Many times, a person who becomes eligible for Medicare declines enrollment in Medicare Part B because he has adequate health benefit coverage as a dependent under his spouse's or his parent's employer sponsored group health plan. At some later time, however, the person may want to enroll in Medicare Part B coverage.

A. When may a Medicare beneficiary, who has declined Part B when first eligible, later enroll in Part B?

Generally, a person who declines Part B when he first becomes eligible, may enroll later only during an open enrollment period, unless the person qualifies for a Special Enrollment Period.

Open enrollment is available only once a year, beginning on January 1 and ending March 31. Coverage is not immediately effective for open enrollees. Part B becomes effective on July 1, following the open enrollment period in which the person is enrolled. If the person does not enroll during the open enrollment period, he must wait for the next open enrollment, unless he qualifies for a Special Enrollment Period.

B. Who qualifies for a Special Enrollment Period?

The Special Enrollment Period is available to some individuals who declined Part B coverage when first eligible because they have other coverage as a dependent of an actively working employee under an employer sponsored group health plan.

A person who has dependent coverage as the spouse of an active employee under an employer sponsored plan will qualify for a special enrollment period when the person loses his dependent coverage. The Special Enrollment Period begins on the date the dependent spouse's coverage ends and continues for eight months. The dependent spouse may enroll in Part B anytime during the eight month Special Enrollment Period .

It is important that the employee (on whom the spouse's dependent insurance coverage is based) be actively working. A dependent spouse will not qualify for a Special Enrollment Period if the other spouse is a retiree, a former employee on a COBRA extension, or has a status other than an actively working employee. A spouse may qualify for a Special Enrollment Period regardless of the number of employees employed by group plan sponsor. However, the number of employees does make a difference for non-spouse dependents who lose their dependent group coverage.

A child (or other non-spouse dependent) who receives dependent coverage based on the active employment of a family member will qualify for a Special Enrollment Period when he loses that coverage, but only when the employer sponsoring the benefit plan has 100 or more employees. If the employer has fewer than 100 employees, the child (or other non-spouse dependent) will not qualify for a Special Enrollment Period and will have to wait for the next open enrollment period to sign up for Part B.

In a nutshell, spouses of active employees employed by an employer of any size and children (or other non-spouse dependents) of active employees employed by an employer with at least 100 employees will qualify for the Special Enrollment Period, if they lose their dependent coverage. The individual may enroll in Part B during the Special Enrollment Period without waiting for the next open enrollment period. ▲

Beth is 46 and has CF. She is an attorney who specializes in disability law. Beth is a Director of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues.

SPIRIT MEDICINE

Enough is Enough

By Isabel Stenzel Byrnes

No matter how full my plate is, writing this "Spirit Medicine" column brings me great satisfaction. But this month, as I started to write, I hit a block. The little voice inside of me starts to talk. *I haven't slept enough to clearly articulate myself. Nor have I enough time to dedicate to this article. Nor do I feel like I have enough ideas.* But herein lies my next topic. I wonder about that concept of "enough." How can we as people–and people with cystic fibrosis (CF)–find a place where we are okay with what is?

I'm pondering the things that I hear routinely from those around me. What do they wish for, personally? I hear people sharing concerns about a lack of money, stuff, looks, health, time… the list goes on and on. In the CF world, those things are often desired, yet lacking in such a real way, because of our life constraints with CF. CF can impose all kinds of feelings of inadequacies. So in this article, I will explore not having enough and having enough, when you have CF.

I often hear statements like the following, When there's… :

Not enough money: Well, haven't you heard that enough?! Many people express that they are lacking money, especially in these slow economic times. I cannot make light of that reality. But, when you have CF, sometimes 'not enough money' is accentuated. It can be hard to work consistently to earn a decent living when your health is a roller coaster. Sometimes we don't have the health to work, and have to live off disability. Sometimes we have to spend a disproportionate amount of money on copayments, food, nutritional supplements, tissue, hygiene products etc. Geez! This is an expensive disease.

Not enough stuff: Welcome to America. We live in a culture that values stuff. We want a bigger house, a bigger car, a bigger TV. My closets are stuffed; I admit I do love to shop for clothes. There is glitz and glamour on TV, in magazines and in the malls, all enticing us to buy more. I don't have an iPod, or iPhone, or iPad… and feel rather out of touch. So many gadgets, so much to want.

Not enough beauty: The media and corporate marketing often imply that we, as Americans, aren't skinny enough, pretty enough, or young enough. Those messages can be extremely damaging to the ego. Though some of us adults with CF (like me) enjoy seeing an occasional wrinkle on our faces, most of us care what we look like and wish to mold into the ideal at some level. For women, CF can make it easier to fit the socially preferred physical model of thinness. For guys, CF can make it harder to be bigger and more muscular. But sickness, including CF, can take away some of the perceived beauty we yearn for—replacing it with paleness and un-pretty things like coughing up mucus and flatulence. It's tough to hide CF sometimes.

> "The cyclical, unpredictable nature of my exacerbations made long-term planning scary and almost hopeless."

ISABEL STENZEL BYRNES

Not enough health: This is a yearning not often heard in mainstream America, because I feel that health isn't always a top priority in our culture. As someone with CF, though, not having enough health was a major burden in my life. I wished to pursue many goals – education, having a relationship and having a family, having a fulfilling career, going on vacation, starting and completing a project, or doing valuable things for others. Often, my illness would rear its ugly head and I would

be limited in some way. The cyclical, unpredictable nature of my exacerbations made long-term planning scary and almost hopeless. I dreamed of having health–and, voila!–all my problems would go away! The 'if-only' of wanting to be healthy was a constant desire that persists today.

Not enough self-care: Many of my CF friends have commented on feeling guilty for not doing enough for their health, amidst life demands like school, work and family. I could always relate to their struggles. I reflect back to when I was fighting CF pre-transplant. I remember being pretty hard on myself, during the days when my health mimicked a boulder slowly rolling down a hill. I often said, "I should do more therapy. I should eat more. I should do more exercise." The list was endless. There was always an overwhelming sense that I wasn't doing enough. This self-blame was its own form of torture, and it wasn't until my disease got really worse that I fully accepted how hard CF was to live with. It wasn't me; CF was just a bi**h.

Not enough time: This is the gold nugget of the CF life. With all my treatments, diabetes care, clinic visits, nap requirements, and the "to-do's" of normal life, I never felt I had enough time to just relax, hang out with friends, and do things I wished to prioritize. On a deeper level, though, I knew what "not enough time" really meant. I was 11 years old and attended CF camp for the first time. At the end of the week, the counselors and kids gathered around a campfire, and read the names of kids who had died the previous year. Though somewhat traumatic, my eyes opened to my own mortality, and I consciously started the stopwatch of my life. I had the urge to soak up all possible possibilities and opportunities in life, filling my plate to the brink– when health permitted.

Shortly before my 30th birthday, I wrote a poem for "Breathing Room" called "Almost." I explored the idea of being almost 30, at a time when I was very, very sick, and shared being almost ready for the end of my life. But, truthfully, I always wanted more time. And I know many of my friends also wanted more time. Surely, their families didn't feel they had enough time with their loved ones.

This list of "not-enough's" could go on and on. Perhaps you've thought of what you'd add, or what you're working on right now. Clearly, there may be many people with CF who feel that there really isn't enough money, stuff, beauty (or, more importantly, the self-confidence it provides,) health, self-care or time. Exploring these briefly does not do justice for the serious impact that "not enough" has on the quality of life of someone with CF. The lack of things in our lives is very real and anxiety-provoking for many.

Now, as I look at the big picture, I think that sometimes our spirits can be dampened by this idea of wanting or yearning for something different, something more and to constantly strive for some level of contentment– out there, somewhere, over there.

I am blessed to be surrounded by wise friends from whom I am constantly learning. A good friend of mine, Alan, came by my house the other day, wiping his face and slumping on the couch, announcing, "Oh, I didn't get enough sleep." Then he stopped himself. "No! I mean… I got the sleep I needed." He proceeded to tell me he is consciously working on ridding the word 'enough' from his vocabulary. I liked his attitude– to catch himself, to reframe his negative judgments to a more accepting place, to be okay with what is.

Wouldn't it be nice for us to be just 'as is?' To reframe our statements like Alan did, and say to ourselves, "I have the money I need. I have the health I need. I'm doing the treatments I can. I have the time I need." Our sense of selves would be so much more secure, if we didn't look for, expect, desire, consume, attain…. more. It takes a great step in awareness to not compare oneself to others (especially healthy others), and to acknowledge you are different from those who have what you want (i.e. wealth, materialism, health, beauty, etc.) This is no simple task. This is hard work.

Could we, even as people with CF, forgive ourselves and accept ourselves for doing enough therapy, for doing enough exercise, for sleeping enough, while managing a full enough plate? Can we respect ourselves for the choices we make to fit it all in? And with regards to time, one of our greatest challenges is to surrender to the fact that we will have lived out our best life within the time given to us. I have known several friends with CF who chose not to pursue lung transplantation. For personal and spiritual reasons, they accepted their natural end, knowing they have lived a full, complete, long-enough life. They have also struggled enough. They let go. It takes dignity and courage to not want more life, and I admire them greatly.

I have also been inspired by the words of Ian Lawton, in his sermon "Abundance and A New Earth." He says, "Abundance is not about getting

Continued on page 11

SPEEDING PAST 50...

Passing Time

By Kathy Russell

Doing nebulizers and other therapies takes up a lot of our time, every day. Do we want to sit and do those treatments and let that time just go by or do we want to find a way to use that time? My preference is to use that time. I find that, usually, I am able to read my e-mail and play some solitaire Mahjongg while I do mine. Keeping busy seems to help the time go faster. I receive enough links to videos and cartoons or jokes to read that I am in a pretty good mood by the time I'm finished.

That is how I spend that time, but that isn't the only time I have to spend in a day. Since I am older, tireder and achier than I used to be and since I am on continuous oxygen, I do not fill my days with very taxing things to do. I find that I am good at sitting in my easy chair, with a heating pad behind my back reading or working puzzles. I get large-print books from the library and can get lost in them. (I love mysteries.) I also like to work jigsaw puzzles. I prefer 1000 piece puzzles of paintings that have blue sky and water in them.

I realize that neither of these activities is very productive, but I have had many years of being productive. I feel that I have earned a chance to be lazy. If I do feel guilty about my sloth, then I use my trusty old laptop computer and do some writing – such as this. Whether that is productive depends on your view. If something that I write helps anyone, then it is productive. If not, then it is a waste of paper. I hope that is more often a plus than a minus.

Being on continuous oxygen can get tiring. It is wonderful to have the oxygen to assist with breathing, but it is somewhat limiting to my activities. It used to be that if I wanted to go out for a walk around our place, I opened the door and went out. Now, if I want to go out, I have to get out my portable concentrator, turn it on, shut down my big concentrator and proceed. Doesn't sound too bad, does it? The portable concentrator has fairly small, hard plastic wheels. They do fine on a solid floor or smooth sidewalk. I live in the country. There are no sidewalks. The ground is uneven and has a lot of hills or rises in it. The wheels don't do so well there.

My husband, being the swell fellow that he is, will carry my concentrator for me. That takes care of the uneven ground, but it just isn't the same as being able to go outside anytime I want. I appreciate his assistance but miss my freedom.

Another time that I notice a lack of freedom is when we are out in our car. It used to be that I could "jump" out of the car and "run" in to a place to pay a bill or return a book. Now, I sit in the car while Paul does the jumping and running. Another freedom lost.

KATHY RUSSELL

Now, lest this turns into nothing but a gripe, I will tell you how great it is to have my portable oxygen concentrator (POC). My POC is small and compact. It fits into the car, next to my legs, without crowding me. I can plug it into the car, while we're on the road, and always have supplemental oxygen. Also, it has a long-life battery that gives me about six hours away from a power source. At home, I keep it plugged into household current, all the time. That way it always is ready to go.

The POC takes very little maintenance. Once each week I wash the air filter and I put on a new cannula. I change my cannula on the big concentrator every week, too, but I change the 50-foot hose on the big concentrator only once-a-month. Both machines keep perking along and are ready at my beck and call.

I have fantasized about what it would be like to have a home that had a high oxygen level so that I could move about with no hose. (As if that could ever happen!) How I would enjoy being untethered once again. Until then, I am so happy that I am able to use supplemental oxygen and still function normally – or as normal as I ever was.

I have written in an earlier issue of *CF Roundtable* that I think of CF as a large cat and I am a mouse that it is toying with. I go along thinking that everything is okay when, all-of-a-sudden, that big ol' cat pounces on me. It slaps me around for a while, and then it lets me go again. …Until the next time… Some of my times away from the cat have been long and happy. Other times, the cat attacks come

frequently and swiftly.

One example of the cat pounces is hemoptysis. All of us who have experienced hemoptysis know how it can appear out of the blue. You can be cruising through life, without a care, when you hear or feel that familiar flutter, rattle or gurgle. You know what it is, before you cough it out. It may be just a few mouthfuls or it may require surgical intervention. No matter the severity, it is darned unsettling. I have been experiencing it for more than 45 years and I still am not used to it. I just don't get as excited by it, anymore.

One problem with coughing up blood is that it is very upsetting to those around you. They are scared for you. They want to do something to assist you, and there usually is nothing that they can do. How frustrating for them. About the most that anyone can do for me is to get me some ice to suck and to keep the tissues handy.

One evening Paul and I were lying in bed, watching a concert on television. I was calm and resting. All at once, I started rattling like crazy. I was really surprised. I thought that maybe it was just some mucus. No such luck. It was another bout of hemoptysis. It didn't last too long and I was able to get a fair night of sleep.

All was fine, until a few days later. I was in Costco, walking at a leisurely pace, when I felt the rattle. I felt real anger toward my CF, because I wasn't doing anything that should cause more hemoptysis. As I continued on my walk, the rattle got more irritating. I was alone and didn't want anyone around me to know what was happening. I managed to get a large enough fold of tissues to catch the blood, as I coughed. I sat on a furniture display and tried to get it to stop. After a few more coughs, I was able to proceed to check-out and find Paul. He got me a big cup of ice and I started holding ice in my mouth. It did the trick. But I still was somewhat unnerved by the incident. I will try to stay nearer to Paul, when I am out like that. I just really don't like it when the cat wins!

It has been a couple of weeks and no more hemoptysis. Now I just have a little pneumonia to keep me alert. That darned cat is always waiting. Fortunately, I caught it early and didn't let it get the best of me. My doc put me on antibiotics that seem to be working. (I am very fortunate because my bugs are pan-sensitive so I can take oral meds and do not have to put up with IVs.) Let's hope that my bugs stay civilized and that oral meds continue to work.

So, time marches on and life goes on. How we use our time is very personal. What works for me may not work at all for you and vice versa. We each will find things to fill our time that can make us feel good about ourselves. I hope that if you have some spare time on your hands you might consider volunteering for USACFA. There always is work to be done to keep the Web site, write the blogs and produce *CF Roundtable*. Think about becoming a part of the group.

Stay healthy and happy. ▲

Kathy is 68 and has CF. She and her husband, Paul, live in Gresham, OR. You may contact her at: krussell@usacfa.org.

Genetic Mutation Information Resource

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

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

The Times They Are A-Changing

By Laura Tillman, President, USACFA

Changes, whether large or small, happen in our lives every year, month, week, and day. Some of them may be for the good, some may not. Whatever life brings, we all must learn to just deal with the hand we're dealt. I'm not trying to be philosophical or sanctimonious here - I'm just pointing out that no matter what, life goes on – even for USACFA.

It doesn't seem possible that another year has passed since USACFA last held elections for the Board of Directors. But, it has, and this year's elections have brought many changes to our organization.

This year we say goodbye to three board members: **Maggie Williamson**, **Cynthia Dunafon**, and **Kathy Russell**.

You may remember Maggie as Maggie Sheehan. Maggie joined our board in 2007 when she was 18 years old! Maggie brought a youthful viewpoint that we certainly miss. However, she had to resign from the board in the winter of 2012 due to a marriage and a move across the country. We wish Maggie all the best in her new adventures in life.

Cynthia Dunafon also felt the need to move on to other things in her life and chose not to run for a director's position. Cynthia became a board member in 2003 and also had a stint as Vice President. Cynthia provided much food for thought for the other board members. She always made contributions to our discussions that were thought-provoking and provocative. As my Vice President, she kept me on my toes! Cynthia will be missed for her creativity and determination in helping USACFA to be the best that it could be.

And, last but not least, we say farewell to Kathy Russell. As most of our readers know, Kathy was a part of USACFA from its beginning in 1990 and elected to the board in 1991. After 22 years of devoting much time and energy to the organization, Kathy felt the need to retire. I wonder why?! There isn't enough room in this column to tell our readers what Kathy has meant to all of us. There were years that it seemed that she single-handedly kept USACFA viable. Kathy has been a role-model, mentor and inspiration to so many. She will be deeply missed as a board member. Luckily for everyone, Kathy is still involved with USACFA as the managing editor of *CF Roundtable* and with her column, "Speeding Past 50". So, it's not really a parting - just a change in function, for which we all are very grateful.

Many believe in the yin and yang of the world so, along with the departures, we have our welcomes! We have three new board members to greet and to whom we extend a warm welcome: **Maria Fioccola**, **Jennifer Hale**, and **Stephanie Rath**. All bring a wonderful enthusiasm as well as unique talents to our board and we look forward to working with them in the coming year. You'll be able to learn more about our new directors in their introductions in this issue of *CF Roundtable*. ▲

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Mailbox

When I was diagnosed in the 1960s, they told my parents to, "Take him home and enjoy him," as I wouldn't be around long. Well, fortunately, they were wrong and, thanks to God, I'm still alive and kicking.

Bob Wright
Pleasant Hill, CA

My name is Audrey. I was diagnosed with CF at birth due to an intestinal blockage and high sweat chloride test results. I have been surviving CF for 33 years now! I live in Virginia and have a great team of doctors that help me achieve the best health I can! I am married to Zach, a wonderful and fully supportive husband. Together we form a Great Strides Walk Team every year in our area to help raise funds for CF research. I still work full time - of which I am very proud. I have a Bachelors degree from Longwood University where I studied psychology. I am active in my church, serving as the clerk there for the last five years. I try to stay as physically active as I can and I enjoy traveling with my husband as much as we can! I continue to live life to the fullest!

Audrey Page
Williamsburg, VA

Regretfully, Sondra (Sandy) Rabinowitz, my wife, passed away. She was the subscriber to *CF Roundtable*. In her memory and in memory of our daughter, who passed away from CF, I will continue to subscribe.

Sidney Rabinowitz
Burke, VA

Thank you so much for continuing to produce such a valuable resource for our CF Community. After all these years, I look forward to what each one of you will write and am always inspired. You're like a group of friends I've never had a chance to meet. Thank you for your hard work!

Wishing all of you much health & happiness,
Dawn McGuinness
Niskayuna, NY

Enclosed is a check for $15 for the renewal of *CF Roundtable* for Jerome Lape. He is an adult male in his 60s now, who was diagnosed in his 40s. His health is declining, although he is very diligent and faithful with his therapies and meds. I am a close friend and want him to continue receiving your newsletter, which he enjoys very much. Thank you!

Stephanie Peterson
Loves Park, IL

Please extend our subscription. We are celebrating Jessica's second transplant anniversary!

Kim Newport (Jessica's mom)
Tampa, FL

STENZEL *continued from page 7*

what you want, or even getting what you think you need. Abundance is about getting what you are. Abundance is liberating. With a spirit of abundance, all that divides us will merge." Having enough, in the psychological and spiritual sense, is having abundance. Abundance is having what you have now.

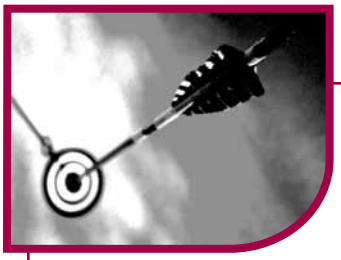
I have no answers for myself on how I can find a place where I believe and accept that enough is enough. So, I have little to offer you. We can try to multi-task. We can try to build a network of support around us. We can try to find community resources to provide practical needs. We can try to balance our wants and needs when tempted. We can make priority lists and do the best we can. We can talk to our health care providers about easing our treatment burden. We can try to simplify, reuse, and buy less. These are just superficial tips on how to manage the "not-enoughs" from the list above. I just try to surround myself with people and art that reminds me of this, each day. So, I end this article with a poem that inspires me to just 'be.'

What We Need is Here – Wendell Berry

Geese appear high over us,
pass, and the sky closes. Abandon,
as in love or sleep, holds
them to their way, clear
in the ancient faith: what we need
is here. And we pray, not
for new earth or heaven, but to be
quiet in heart, and in eye,
clear. What we need is here.

From: "The Wild Geese," by Wendell Berry, Selected Poems, 1998 ▲

Isabel is 40 and has CF. She and her husband, Andrew, live in Redwood City, CA. You may contact her at: Isabel@usacfa.org.



FOCUS TOPIC

RESPIRATORY THERAPY ACTIVITIES

The Treatment Game

By Mark Levine

As Julie Desch said in the last *CF Roundtable*, "the one perfect workout is the one you will do." I take a similar approach when planning my treatments. I have to admit, that I'm not big on chores and I certainly do not look forward to things that I do not like to do. Who does? I consider doing my treatments a game, not a chore. And like any game you play, the mental aspect is key, but they could certainly be fun to play.

To play the treatment game, you have to make a few rules. The most important rule I have setup for myself is this - never miss a treatment. I do two hypertonic saline treatments and one Pulmozyme® treatment per day. Every other month, I go on Cayston® and do three treatments of those per day. That means I am often doing six treatments each day and, from an outsider's perspective, that may seem daunting. But the thing is, as soon as it seems daunting to you, you've lost. The object is to make it not ever seem daunting.

So here are some suggestions.

Suggestion number one is – Treat yourself to a treatment. Doing something you enjoy while doing your treatments makes it seem like a bonus. Sometimes I will watch TV, sometimes I will surf the internet, while other times I will play a game or read a book (if I'm not doing *The Vest*® at the same time). Often I will work my stay-at-home business, which requires computer work, while doing my treatments and, in that light, I plan for it and look forward to it.

Multitasking is a concept I try to exploit while doing treatments. Each morning on the way to my workout at the gym, I do my treatments in the car using my PARI-Trek® plugged into the 12V outlet. My workout is awesome for helping me clear my chest and the treatments right before prove quite effective.

One of my favorite setups can be found in my basement where I do a breathing treatment while using the Vest working out on the elliptical trainer and watching a recorded Daily Show with Jon Stewart or Colbert Report. It truly is a treat. I've done the same routine while also on IV antibiotics. That is five activities at once and I have to admit that I'm quite proud of that arrangement.

NEBULIZING DURING A WHITE WATER RAFTING TRIP IN UTAH, WHERE THERE WAS NO ELECTRICITY.

Even though I can do the above in the comfort of my own home, when I travel, I try to get out of my room to do my treatments. Once again, using my Pari Trek, which is the perfect travel size air compressor, I head to the gym and do my treatment while riding a stationary bike, walking on a treadmill or using an elliptical trainer. I always bring an extension cord just in case the closest plug is not close enough. And of course an iPod makes the experience that much more enjoyable. Perhaps you're asking yourself, "And what about those other people in the gym who may look at you funny?" Let 'em look. It's really not much to look at for more than about 15 seconds. Then, as to be expected, my doing a treatment on a treadmill takes a backseat to whatever was keeping them occupied before I began.

Finally, three Cayston treatments a day turns out to be very doable although, at first glance, it could seem like a handful. Of course, at three to four minutes per treatment, it is easy to sneak in almost anytime, anywhere. Here's how I pull it off. I do my first treatment of the day after my workout. I clean the special Altera nebulizer with soap and water and bring it up to dry on a paper towel in my office. Then I put the mid-day Cayston treatment on my Outlook schedule and simply do the quick and silent treatment right at my desk. As soon as I get home, I sterilize all of my nebulizers and set everything out to dry before finishing my last treatment just before bed.

Bottom line is this. With a little planning, foresight and optimism, you can do it all, have fun and get a lot of things done at the same time. Play the treatment game and you'll be happier all around. Happy breathing. ▲

Mark is 42 and has CF. He is a Director of USACFA and his contact information is on page 2.

noons a week or that I sleep 9-10 hours a night, they don't understand and some even make jokes about it. Then I get hurt. This isn't fair to them, of course, because they see me only as my public "healthy" self. And so I continue to struggle with how to handle this group.

Overall, I am still a healthy world wannabe. I have accepted my disease and am fully committed to doing all my treatments and taking care of myself but still try to fit in to the healthy world whenever possible. My motto is that I live as if I don't have CF but treat my body knowing that I do. I don't know if this is a partial denial or a good way to look at it but, so far, it works pretty well for me.

Then there is *The CF World*. The CF world has been a very bizarre place for me because the severity of it in each person varies widely. When I was diagnosed in 1972, I was 12 months old and the average life expectancy for a child with CF was 10. Because of this, my parents were extremely overprotective of me - they didn't like me going to sleepovers and every year we went to Florida for a month to get away from the main flu and cold season in Connecticut. I remember being aware of having CF and having to do nebulizer treatments when I got a cold, but overall, I didn't feel sick. I had to take enzymes and my mom and I had to get up extra early each school morning so she could do hand clapping therapy on my back, but I never coughed up much.

By the time I hit my teens, my mom was working full time and I had gotten a weird machine that was supposed to replace her clapping therapy. I decided that the therapy was stupid, that I wasn't really sick, and that I didn't need to do it. The only thing I did for my CF was take enzymes to help with my digestion, due to embarrassing consequences if I didn't.

Fast forward to age 22 - I was living in Michigan, had a terrible scary cough and was coughing up blood. I set an appointment with a brand new doctor, since I had just moved there. I remember being asked all kinds of bizarre questions like - did my stools sink or float. (WHAT? Who looks at it? And why do you care?) I had a battery of tests run on me including the standard PFT. I was totally unaware of my numbers and didn't know what they meant. I'll never forget the moment my doctor looked me right in the eyes and said, "If you keep doing this little to treat your CF, you will be dead by the time you're 26."

Twenty-six? I was devastated. Somehow, in the back of my head, I had decided that I would probably die early because of my CF – like maybe in my 60s - but 26? He immediately put me in the hospital for my first "clean out", had me drink a GALLON of GoLytely® (literally the worst stuff I have ever tasted!), get an enema, take 47 black chalk pills and then test my bowels and did a variety of other unpleasant, humiliating tests. I was given books and pamphlets about CF. I met with the entire «CF team» (I had never heard of such a thing!) including a nurse specialist, a respiratory therapist, a dietician, a therapist and a social worker. When I went home I was given Albuterol and Pulmozyme to nebulize twice a day and, soon after, acquired the brand new «vest» machine. Welcome to the real world of CF, Jen.

During my hospital stay of two weeks, there were many others on the same floor of the hospital as I was, who were going through their clean outs – most of them children and teens. Of course we were not allowed to hang out or go into each others' rooms, but word spreads, people spy on each other and nurses talk. After being there for two days, I was informed that I had received a nickname from the other people with CF. I was called «the bitch.» This was not due to my personality, but rather it was due to the fact that I did not need my first clean out until age 22. They despised me for being so healthy. Healthy? I just received the most devastating news of my life about the severity of my illness - and they were upset because I was so healthy?

I still continue to live out this dichotomy today. I had to retire at age 34, because regular work hours were getting to be too much of a drain on me (I work 8 hours a week now). Of course I do my two hours of breathing treatments a day and am on so many medicines that all of the people who work at the pharmacy know me by name. But to many in the CF world, I am doing well. I am a surprise to doctors and nurses. (I can't tell you how many times I have had my lungs listened to, only to be told how «great» I sound – I have only 40% lung function and you tell me I sound great?) For some who have CF, or have children with CF, I am a vision of hope; for others I am secretly hated for being so lucky.

Since "joining" the CF world, I have gotten involved in some great CF organizations and met some of the most amazing people in my life. Many of them needed lung transplants in their 20s and many of them are even older and healthier than I am. No matter what their health status is, I have found that most CF people are resilient, are fighters and are givers. They are caring and will share in your heartaches as well as your joys. They will cheer when your Pulmonary Function Tests are up and empathize when you have to go into the hospital. I feel so fortunate to have met so many incredible people.

Right now the average life expectancy for someone with CF is 37 and I am 41. Compared to many who have suffered so much with CF, I am fortunate. Compared to most people who live in the healthy world, I was born with a great burden. In the end, I think they both are right. ▲

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

Meet the New Directors

Maria Fiocolla

I am a people-person. I was born, surrounded by loved ones, in September of 1984, and I continue to surround myself with my loved ones. I was diagnosed with cystic fibrosis (CF) at six months of age and still stand strong at 27 years of age. I am a middle child, the only girl and only member in the family to have CF. I have not had a transplant. I have been blessed with the doctors at Northwestern Memorial Hospital's Cystic Fibrosis Clinic, since 2005. Prior to this clinic I was untreated. I am currently unemployed but, no worries, I keep busy. As newly elected to the USACFA Board of Directors, I am ecstatic about volunteering my time to the CF community. I was also elected Secretary for the Northwestern University Patient Advisory Board earlier this year. Other than my CF involvement, I love spending time with my family and friends. I reside in Gurnee, Illinois with my fiancé and his two boys. My fiancé, Matthew, has been a great supporter and inspiration. His love of welding has inspired me to share my love of writing with others. I now have a self-published poetry book titled "Sincerely Yours" at: www.blurb.com. The profits of this book are sent to the Cystic Fibrosis Foundation. His two boys reside with us and really lightened up my world as well. Alexei is 14 years old and going into high school this autumn. Nikolai is 9 years old and he will be in the 4th grade. I look forward to saying they are my stepsons, since I do not have children of my own. We do have two four-legged family members. Shadow (female) and Dali (male) are the family cats. This is just a glimpse of my life, my family and my inspiration. Thank you for reading!! ▲

MARIA FIOCCOLA

Jennifer Hale

Hello *CF Roundtable* readers. I am Jennifer Hale. I hope you have been enjoying my column "Coughing With A Smile". I am proud and honored to say I am now one of the new directors of USACFA! A little background about me: I was diagnosed with CF at the age of two and I am proud to say I am now 40 with CF and CFRD. I spent most of my life in the suburbs of Chicago and now live in sunny Florida with my wonderful husband to whom I have been married for 15 years. We love the weather and all the available outdoor activities here in Florida. I went to college at Eastern Illinois University and went on to work full time for a few years until having to take an early medical retirement. Now my days are filled with caring for myself, my health and my husband. I enjoy being active and love the beach. My FEV_1 has decreased, but I have just modified my lifestyle and activity to adjust. The latest adjustment for me has been my portable oxygen tank that I use while exercising in the local gym. Nothing will stop me. Keep on keeping on! I look forward to hearing from the readers of *CF Roundtable* and I hope to write inspiring, motivating and funny articles. Thank you. ▲

JENNIFER HALE

Meet the New Directors *Continued*

Stephanie Rath

Hi! My name is Stephanie Devine Rath. I'm one of the newbies to the USACFA board. I'm really excited to be part of this organization. I've been reading CF Roundtable for more years than I can remember and it has always been a source of inspiration and good information.

I was diagnosed with CF at age 19 via genetic testing. Sweat chloride tests prior to that age were inconclusive. I was almost transplanted at age 23 but was able to rehabilitate myself via exercise and strict adherence to therapies. Currently I'm 43 years old and loving every minute. It's not always been easy but most of my life has been a wonderful adventure. My husband, Randy, & I love to travel, see live music, and enjoy time at home with our Boston terriers, Rocky & Rosie. I also enjoy yoga, gardening, good food and I am learning to sew.

In the last few years my health has declined. I retired from my position as Tax Manager for Ernst & Young in 2009. Since then I've been through one transplant workup and treatment for rectal cancer. I will be eligible for transplant again in the fall of this year. Currently things are stable and I'm on oxygen only at night and with exercise. My determination and a positive attitude have helped me through the many challenges of CF and I hope to be overcoming them for many years to come.

STEPHANIE RATH

I stay involved in the CF community locally and online. For the last 14 years, I've chaired the Festiv-ale Beer tasting for the Indiana CF Foundation chapter. I speak at that event and whenever needed to help spread the word about CF. My other interests include sharing my experiences via blogging, CF web forums, and Facebook groups. I'm honored to be selected to serve on the board and have the opportunity to be even more involved in the CF community. ▲

Information from the Internet...

Compiled by Laura Tillman

This issue brings a potpourri of articles from the Internet

Press Releases

Should Cystic Fibrosis Patients Get So Many Antibiotics?

Study suggests that less-aggressive antibiotic therapy might be better

Antibiotics can prolong cystic fibrosis patients' lives, but the drugs also help treatment-resistant bacteria thrive in their lungs, a new, small study suggests. The findings from the 10-year investigation suggest, but do not prove, that the current standard of aggressive antibiotic treatment for cystic fibrosis patients may not always be the best approach. It's common to use antibiotics to control infection in cystic fibrosis patients' lungs, but maintaining a more diverse range of bacteria in the lungs may help some patients stay healthy longer. Aggressive use of antibiotics actually lowers the diversity of lung bacteria, resulting in infections that are increasingly difficult to treat. A more diverse community of lung bacteria may help keep the most dangerous strains in check.

http://tinyurl.com/7d9o8st

Research Shows Why One Bacterial Infection is so Deadly in Cystic Fibrosis

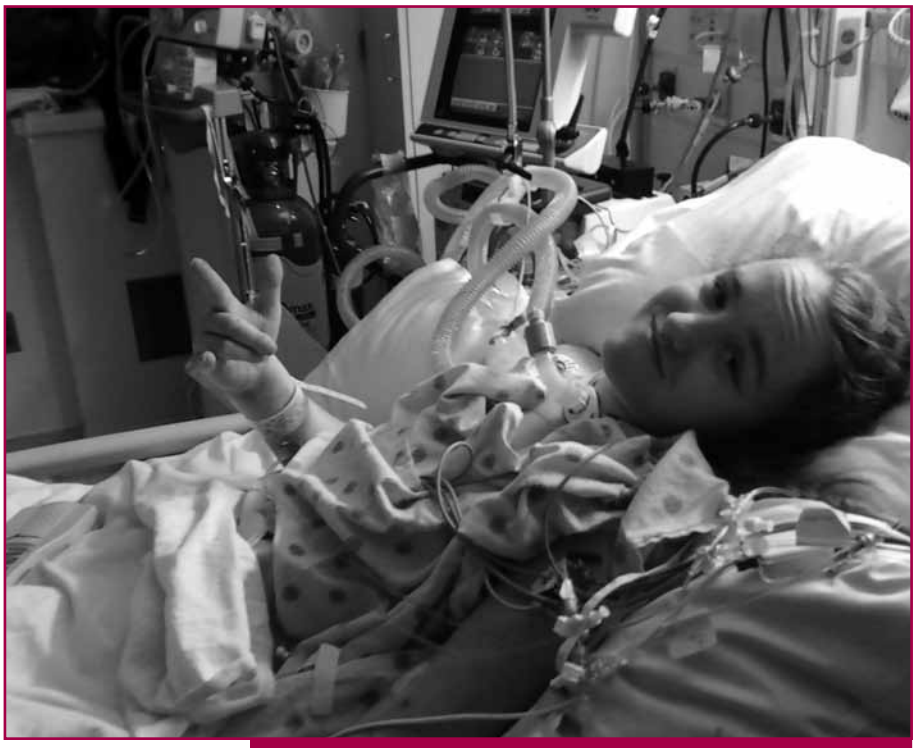
Burkholderia cenocepacia, causes a severe and persistent lung infection in patients with CF and is resistant to nearly all known antibiotics. The Ohio State University researchers have

Continued on page 18

Who's Afraid of the Man in the Pale Blue Bunny Slippers?

By Annie Opatz

Children carry within them the most intimate life secrets: the perfect sprinkle-to-cupcake ratio; every ounce of muscle memory hidden in Hannah Montana's "Hoedown Throwdown" dance; the truth of stepping on a crack and breaking a back; and, even if it seems geometrically impossible, your little sister can fit down the laundry chute. With all of this crucial information swimming within a child, how can there be any room for what adults call "The Big Things In Life"? The birds and the bees, what I want to be when I grow up, vegetables, divorce, cleaning my room, where does it all fit in? And what about that thing that no one ever seems to talk about -- that thing clouded in muted, gray secrecy? It's the farm where my first dog Muggzie ended up, the morning I saw my father cry for the first time, the one day when all those kids went to school but never came back, you know, somewhere out in Colorado.



ANNIE OPATZ RECOVERING SHORTLY AFTER HER BILATERAL LUNG TRANSPLANT.

What about death? Where does death fit in a child's life, especially a child living with cystic fibrosis?

For me, death is stored in the deepest, darkest crevices of my DNA. As if it was already predestined; the result of a heated game of chess between God and Darwin and I am its outcome. Understanding death has become a part of me, surrounding me, following and eluding me, like Peter Pan's shadow, always there but never captured. Playing this cat and mouse game with death has always been a part of living with cystic fibrosis. More than pills, treatments or hospital visits, greeting death with a tenacious smile has become routine. As I have grown older, some of my friends have even lightheartedly asked me, "Was the grim reaper your imaginary friend growing up?" And the truth is that, for every person living with cystic fibrosis, he is. Because living with cystic fibrosis means living with the constant reminder that the most meaningful life and the most heartbreaking death may be only a breath away.

When I was a child, death and I would play together in the sandbox, sharing stories but few laughs. I would tell him how I was feeling and of my immortal fears. He would sit in silence, watching me sift sand through my hands. As I grew older, he rode on the backseat of my tandem bicycle, his sickle poking me in the back. He was my first passenger the day I passed my driver's test; he sat next to me on that cold, rainy November day when I received the call letting me know I was finally placed on the lung transplant list, and he is here even now, a year and a half after I was blessed to receive a brand new set of lungs (he's in the other room, watching *Survivor*… go figure.)

Unfortunately, for the cystic fibrosis community, death is not the imaginary friend standing in the corner eating a peanut butter and banana sandwich. It is very real and it is inevitable. Despite many remarkable breakthroughs in medical care and treatment, the median life expectancy for a person living with cystic

fibrosis in the United States is 37.4 years. And for those living in other countries it is as low as 20.

So what does this mean for me, a 25-year-old sunny California college student just trying to get by? It means knowing the difference between understanding and accepting what my future holds. It means walking that ever so thin tightrope between feeling sorry for and grieving for myself and my illness. Most importantly it means allowing others to learn from both my strengths and weaknesses as a person living and still breathing with CF.

Now I wish I could sit here and say I am not afraid of death. I wish I could say I will gladly go with him when he asks me to go, in the middle of the night and toilet paper the pearly gates. But the truth is, despite his constant companionship, I am afraid of death. Like in so many Hollywood movies, I envision a tall, sinister figure, shrouded in a black mantle. For the most part he is silent, except for every few seconds when he glances at his watch and whispers, "I'll be right back," escaping into thin air, going off to do what he does best, only to return a few seconds later, brushing the dust off his shoulders.

It is safe to say death has always terrified me. That is until a little over a year ago, when his hollow, blackened face began to change. I began to question who he was and I finally had to ask him, "Are you afraid, too?" Not only is death afraid, is the potential cause of my death afraid? Is cystic fibrosis afraid?

In July of 2009 an acquaintance of mine lost her lifelong battle with cystic fibrosis. We were not extremely good friends. I followed her blog religiously and wrote to her often to let her know how much I appreciated her candor and positive attitude, despite the unforgiving progression of the disease. But when she died, it hit me particularly hard. It wasn't just that it was cystic fibrosis and I have cystic fibrosis, it wasn't just that she was 26 and I was 23, it honestly felt like the appearance of death was purposeful and resolute. Like death itself was childishly sticking his tongue out at me saying, "Naa naa naa naa I took your friend!!" Although I cried for days, I still felt as if death was mocking me.

> "Playing this cat and mouse game with death has always been a part of living with cystic fibrosis."

It was within this shell of grief that I began to question my friend's death. Not in the sense of why did she have to die, why do I have to die, why cystic fibrosis at all; but more in the sense of who are you? I couldn't help but ask, "Death, do you like what you do? Are you proud of this disease? Does it make your job easier?" This shadowy figure, which has been genetically forced into my every day existence, began to melt away and I began to see death not as this grandiose and mysterious form, rather like a child who holds the same intimate truths we do as children, the truths of cupcakes and laundry chutes just snug enough to slide down. I began to strip away the façade of death and what my relationship with death and cystic fibrosis really meant as well as what my own terminality will be.

My death is different; he is eclectic. My death rode that old tandem bicycle in pastel blue bunny slippers, uses his sickle as a light-saber, enjoys eating bowls of Captain Crunch in the bathtub as much as I do and is haunted by the humans he has taken more than we are haunted by him. With this change in my outlook on death also came a change in my outlook of cystic fibrosis. My cystic fibrosis wavers between flip flops and four inch heels, eats copious amounts of bacon, listens to both the Neil Young and Lady Gaga and befriended a fellow medical anthropologist, who would inevitably become my transplant surgeon.

So how does this change me; how does this change my death? More importantly what does this mean for me going forward, living with CF? This new death does not change my story. I still take pills, I still do treatments, I still open my favorite CF website (www.cysticfibrosis.com) every day and read the posts, "another angel has gotten their wings, breathe easy". I still have an illness to which I will eventually succumb. What it does is this: it takes the fear away. It allows my own personal anxiety to be stripped away, leaving a temperate relationship with death, one based on a mutual respect and understanding for one another. I respect that he unfortunately has a job to do and he understands that myself along with all of my fellow CFers will never come easy. ▲

Annie is 25 and has CF. She lives in Rohnert Park, CA and is a student at Sonoma State University.

determined that *B. cenocepacia* bacteria interfere with an important survival process in cells whose job is to fight infection. This phenomenon is even stronger in CF patients, so the infection exacerbates the cell malfunction. The research group also showed that rapamycin, an existing drug known to stimulate this cell-survival process, called autophagy, helped control B. cenocepacia infection in mice that serve as a model for cystic fibrosis. These cells that can use autophagy to clear infection are the macrophages, which are first-responders in the immune system that essentially eat offending pathogens. The scientists also dissected the role of a molecule called p62, which plays a role in the autophagy process. They found that p62 inside macrophages is influential in controlling *B. cenocepacia* infection.
http://tinyurl.com/7vkyl8u

Phage therapy can combat Pseudomonas infections in CF patients

Researchers took advantage of a method called phage therapy, which has been practiced in Eastern Europe for decades but has received relatively little attention in the West. This involves identifying and characterizing bacterial viruses [phage] which can attack and kill Pseudomonas aeruginosa within minutes of initial contact. The phage ‹infect› the infecting bacteria and multiply within them, leading to cell death and the release of hundreds of new phage. One of the advantages of phage therapy is that any viruses which ‹find› a target multiply at the target site, generating more viruses and amplifying the therapeutic effect. The researchers showed that their newly isolated phages could very effectively kill Pseudomonas aeruginosa in situations mimicking real infections, and could eliminate all strains of Pseudomonas isolated from Cystic fibrosis patients.
http://tinyurl.com/795kwsh

Largest gene therapy trial for cystic fibrosis begins

A consortium of researchers from Oxford University, Imperial College London and the University of Edinburgh will begin a mid-stage trial later this month for a cystic fibrosis gene therapy delivered to the lungs with a nebulizer. They'll also begin a laboratory trial using a more advanced version of the same drug designed to ride in a modified virus to reach its target. The phase two clinical trial will involve 130 cystic fibrosis patients ages 12 and older using an inhaler to breathe in a working copy of the cystic fibrosis gene once a month for a year.
http://tinyurl.com/7l5r6vl

Researchers Studying Defective Protein in Search of Cystic Fibrosis Treatment

New research at Ryerson University, in partnership with The Hospital for Sick Children, is examining the interaction between lab-tested drugs and the defective protein that causes CF to understand how and why the drugs work and to create stronger, more powerful second generation drugs to treat CF. Researchers are examining the structures of failed drugs that, in a previous lab setting fixed the defective protein but, for a variety of reasons, never made it to patient trials. The team modifies the structure of the drug slightly and in such a way that biochemists can examine how each of the failed drugs interacts with the defective protein.
http://tinyurl.com/7xnlvy3

Boston scientists grow lung tissue from cystic fibrosis patients' skin cells

Two teams of Boston scientists have developed new ways to turn stem cells into different types of lung tissue. Beginning with the skin cells of patients with CF, Jayaraj Rajagopal and his colleagues at Massachusetts General Hospital (MGH) first created induced pluripotent stem (iPS) cells, and then used those cells to create human disease-specific functioning lung epithelium, the tissue that lines the airways and is the site of the most lethal aspect of CF, where the genes cause irreversible lung disease and inexorable respiratory failure. That tissue, which researchers now can grow

Continued on page 25

Vrksasana

Magnificent tree
reaching upward toward the sky
grounding me

Miraculous breath
flowing throughout my body
sustaining me

Mind, body, spirit
seeking balance and wholeness
strengthening me

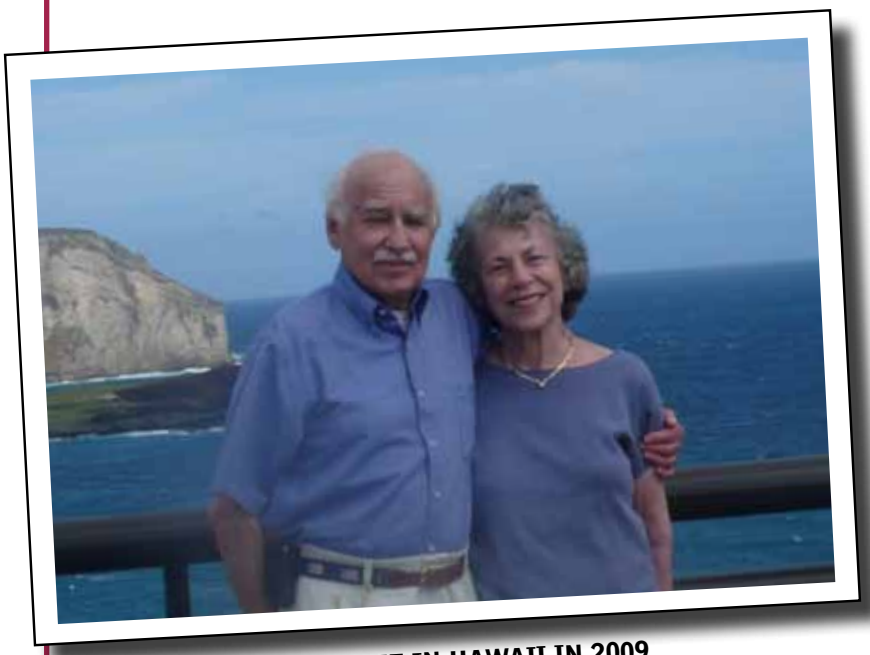
-R. Petras, 2000

PHOTO BY STEPHEN BOYER

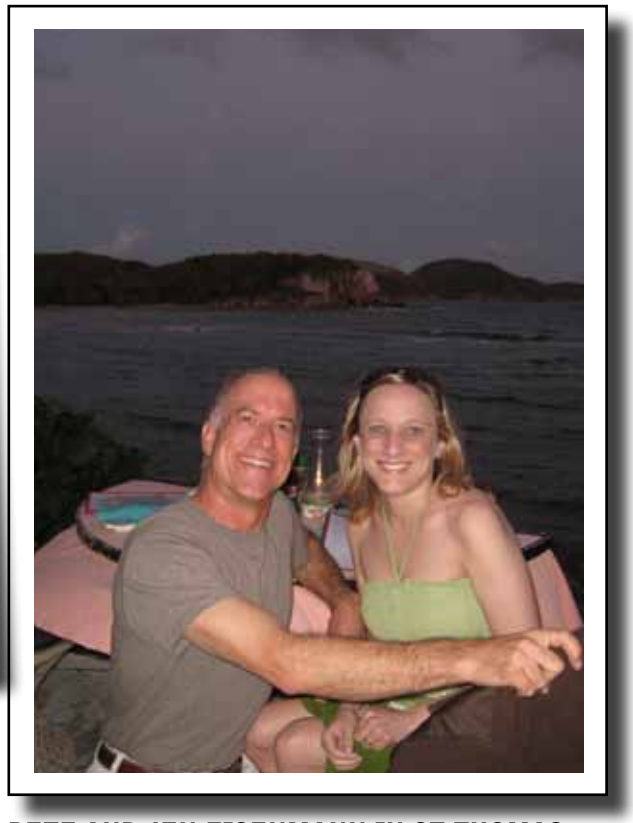
"Through the Looking Glass: Images of Adults with Cystic Fibrosis" and "Caregiver Stories" are projects of Breathing Room, a non-profit organization. Breathing Room hosts these and other projects to facilitate open and candid communication in the CF community, supports the development of a community 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...

LEFT TO RIGHT: JOELLE, ADAM, MARK AND BROOKE LEVINE AT THIS YEAR'S CF WALK.

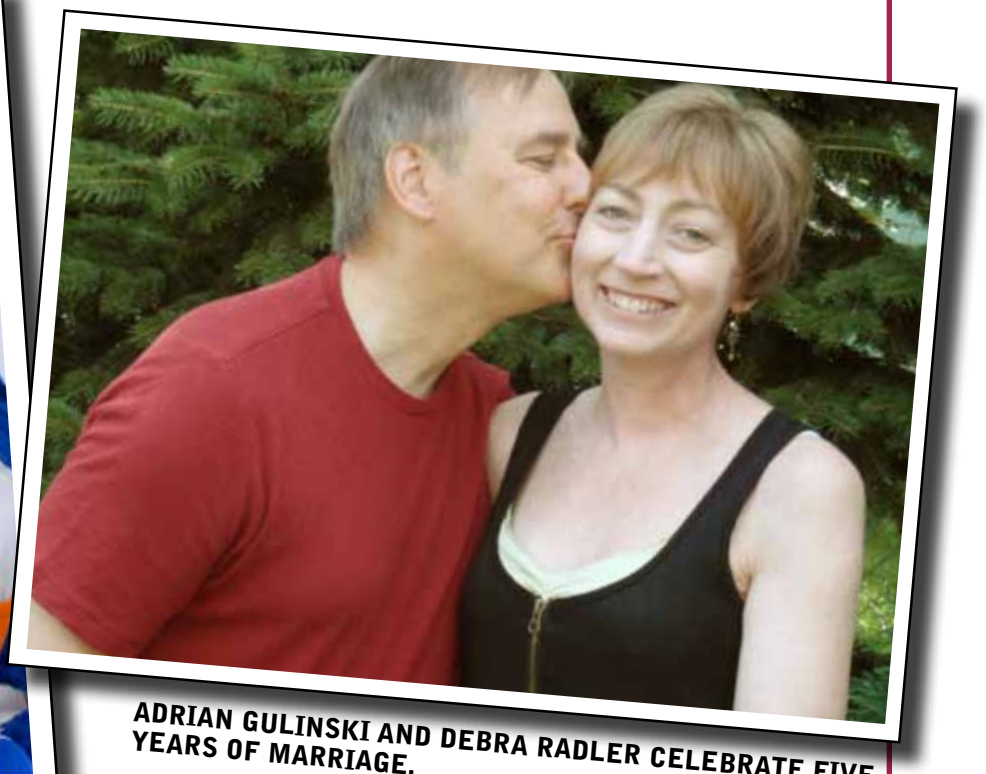


HAL AND PHYLLIS SOLOFF IN HAWAII IN 2009.



PETE AND JEN EISENMANN IN ST. THOMAS. US VIRGIN ISLANDS.

JORDAN HERSKOWITZ IN COSTUME FOR HIS ONE-MAN SHOW,"GROWING UP JORDY PORDY."



ADRIAN GULINSKI AND DEBRA RADLER CELEBRATE FIVE YEARS OF MARRIAGE.

LEFT TO RIGHT: KATHY AND PAUL RUSSELL, ANDREA EISENMAN AND LEW AND LAURA TILLMAN AT THE MOUNTAIN WEST CF CONFERENCE IN LAS VEGAS, NEVADA.

BOOK REVIEW

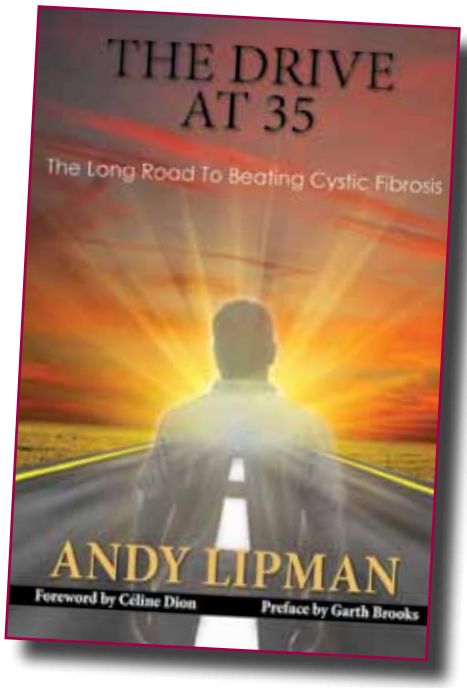
The Drive at 35

The Long Road To Beating Cystic Fibrosis

By Andy Lipman
Reviewed by Paul Feld

The Drive at 35… takes a look at one man's attempt to beat CF while working full-time, being married to a wife with MS, having two children, being a key fundraiser, and trying to stay positive and a fighter throughout. This 234 page book chronicles Andy's life from the time of diagnosis right after birth to present day. As far as 'life with CF' challenges goes, I'd say Andy has had a fairly normal life for a person with cystic fibrosis. He has endured thousands of hours of therapy, typical CF roadblocks such as intestinal issues and depression, and several family issues. At points in his book, I got the impression that family, friends, and various college buddies often put the CF issues in the background and made the personal issues more challenging for him.

The inspirational aspects of the book proved to me that I have the hope that Andy will be writing 'Part 3' of this book when he turns 55.



Andy writes about the older sister he never met, who died from CF at just two weeks of age. He talks about his dreams about her, and how he firmly believes all the influences she has had on him in his life. He turned his story about his sister Wendy into an amazing year-after-year fundraising event to support the CF community: *A Wish for Wendy Foundation*.

Andy is now 38, and has just passed the median life expectancy of those born with CF. The saddest parts of his book are the friends and family he has lost along the way, while he continues to beat CF and live on. It just proves that life has its challenges for everyone, and no one knows when his or her life might end.

I enjoyed reading about Andy and most of his life experiences to date. I hope he continues to overcome the obstacles CF throws at him, and that he will continue to share his life story down the road. Andy's two books are available through the website http://www.andylipman.org/books.cfm, and this particular edition is $14.99, with $5 donated to the Cystic Fibrosis Foundation.

I highly recommend this book to young adults with CF, and the inspiration and character buildup it should provide. Those of us 'older' CF adults can read and reminisce about times gone by, and play the game of predicting what still lies ahead for Andy. ▲

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!

Voices from the Roundtable

What Can You Learn in 81 Years?

By Hal Soloff

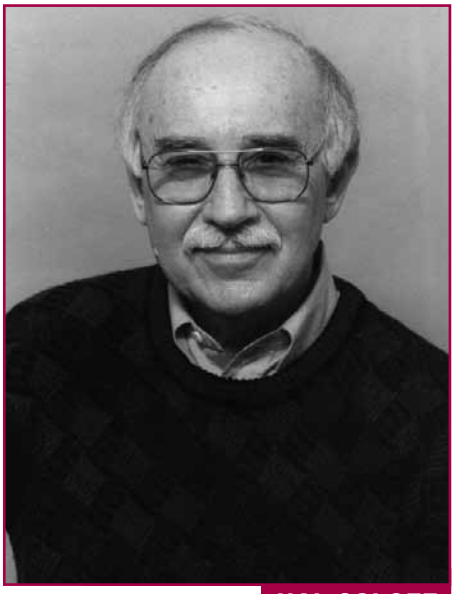
When I was born, from what my mother later told me, I was bright red, jaundiced and crying in distress. When she took me home the next evening, I was still red and crying. She picked up the telephone receiver and asked the operator, as there were no dials in those days, to get the doctor for me. She knew where the doctor was playing cards. He came quickly, recognized the problem, and flushed me out. It's called meconium ileus.

When I was eight we moved to Dallas, Texas, where my sister was born. I spent the summer in the hospital on IVs, as I couldn't tolerate the heat. Patches of salt would form on my forehead, but the child psychologist in the hospital said my illness was because I was jealous of my baby sister. Other than the summer in Dallas, my mother always insisted that she take me, and later my sister, to Connecticut to be near the shore. I loved walking the beach and breathing the salt air.

We moved to Chattanooga, Tennessee, when I was twelve. My digestive problems and bowel movements were attributed to my being "nervous". The junk I was coughing up was diagnosed as from "post nasal drip". I was put on sulfa drugs, as my sinuses were constantly infected. The polyps were taken out by a snare. I was diagnosed as having "allergies". I was underweight as I had been put on a diet due to the allergies.

At age ten, I had learned how to play the drums and, when I was in high school, I played in the band and orchestra, and played the timpani in the local symphony orchestra. It was excellent exercise. I joined the boxing team, and I fought in the Golden Gloves. I also won many blue ribbon medals in band drum competitions. At seventeen, I went to the Juilliard School of Music pre-college program in New York.

I had my first Caldwell-Luc sinus surgery just before I went to Northwestern University, at age eighteen. I was in the Northwestern marching band. In my second year of college, after my second Caldwell-Luc, a war had started in Korea. I went to the Air Force recruiting office, thinking they'd never take me.



HAL SOLOFF

By the sergeant putting his foot on the scale, and the doctor giving me a superficial physical, they took me. My Air Force experience lasted two years. When we were flying, the oxygen failed, and my sinuses let loose with a lot of blood. Others had their ear drums ruptured, so they flew us to an Air Force hospital, and after three weeks of TLC, I was discharged as a disabled veteran. I had three years of the G.I. Bill, which helped me get through law school.

In my second year of law school, between semesters, I visited my mother's family in Connecticut. It was a fortuitous visit, as I met my wife to be, Phyllis. How I was able to convince the most intelligent and beautiful girl I had ever met to marry me, I'll never learn. She probably felt sorry for the runt of the litter.

I got my first job as an attorney with a New York Stock Exchange firm in Springfield, Massachusetts. I also became a Registered Representative of the Exchange. I was still coughing, and growing polyps, so I decided we should drive to Boston, to see the ENT specialist I had seen as a child, Dr. Carlyl Flake, at Children's Hospital. After checking the mess in my sinuses, Dr. Flake said he wanted to introduce me to another doctor, and I met Harry Shwachman, M.D. Dr. Shwachman asked to give me some tests. My left arm had electrodes attached and it was wrapped in opaque plastic. The electricity was turned on, and I felt light shocks that didn't cause pain or capital punishment. After he unwrapped my arm, he collected the sweat and measured the salt content. Dr. Shwachman then made the diagnosis – I had cystic fibrosis (CF).

With Phyllis present, he explained the disease. He brought out a petri dish with a maroon colored mass, and told me to put the fingers of my hand on it. Where I touched the mass, it turned white. He nodded, as if it confirmed the original diagnosis. He listened to my chest, front and back, and mumbled about wheezing and crackles. He diagnosed my sister two weeks later, but it was too late for her, and she died at age twenty from the ravages of CF. It was the first tragedy in my life.

Dr. Shwachman prescribed many

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medications, including antibiotics and enzymes. He gave Phyllis a bottle of oil for her to cook with, and showed her how to give me physical therapy to loosen the mucous. We drove to Boston frequently, and I would have routine salt tests, X-rays now and then, sputum cultures and long discussions with Dr. Shwachman. Phyllis was always there and, most often, took notes to ensure I would follow through on his suggestions. My local doctor would call Dr. Shwachman for consultations.

In 1961, Dr. Shwachman asked me to volunteer to be the first live CF male to have vas deferens exploratory surgery. I agreed. The surgery was performed by Dr. Robert Hepburn, Katherine Hepburn's brother. The results were published in the "New England Journal of Medicine." The surgery had affirmed that almost all males with CF have either no or an undeveloped vas deferens.

In 1962, Dr. Shwachman, after listening to my lungs and showing us my X-rays, said that my lungs were deteriorating quickly and I should consider a less arduous lifestyle or he didn't think I could go on for more than two or three years. On the two hour trip home, I got a stern lecture from my wife, that I should give up law and securities and consider teaching, since it would give me less stress and more time to do the routines necessary to survive CF. I went into New York and tendered my resignation to the Chairman of the firm. My law career ended and I began my career in teaching.

My career as a teacher lasted for twenty seven years. I did have IVs more frequently. At first I had them at the local hospital, but my primary care doctor, due to concerns about infections, decided it would be best if I did them at home. I began a series of antibiotics and mucous thinning inhalations through the years. I also began using a hand held machine to pound my chest.

In 1965, we adopted David, our first child. When David was two, in 1967, we decided to travel to Europe for seven weeks. We couldn't put David in a kennel (it's a joke) or leave him with Phyllis's parents, so we took him along. We flew to Gothenburg, Sweden, picked up a Volvo at the factory and drove to Oslo. After three days, we drove north above the tundra line on single lane dirt roads, staying in small towns along the way. I had electrical converters to use my small inhalation machine. We were high above the fjords and the scenery was spectacular.

We then drove south to Bergen. From Bergen we put the car on what was called a packet (mail) boat, the "Arundel". The boat stopped at most villages along the coast of Norway, past fjords, on the way to Oslo. Back in Oslo, we drove to Sweden, and then over to Denmark. We stayed in Copenhagen and learned to get a student from the university to help us with David. From Denmark, we took the ferry to Puttgarden, Germany. We drove to Holland, and stopped in Groningen. Amsterdam was a short drive away and we enjoyed Amsterdam and the museums and canals.

From Amsterdam, we drove to Brussels, spent a couple of days there, and saw the beautiful Old City. We drove to Luxembourg, through northern France to Fontainebleau for a day, then Paris. The "Mona Lisa" at the Louvre was a surprise, as it is relatively small. We had lunch at the Tour Eiffel and took a day trip to Versailles. We drove to Dunkirk three days later after touring Paris, and took the ferry across the English Channel to Britain. We stayed at a B &B on the small island of Sheerness, at the mouth of the Thames.

When we got to London, we stayed at a very good small hotel, and as a joke, since it was a rainy day and we were bored, Phyllis suggested that I look for relatives in the phone book. I did and it was there that we established our relationship with cousins I hadn't known existed. We now have been a family for forty-five years, including family reunions with Australian cousins, weddings and just getting together.

In 1968, we adopted our second child, Jennifer. We now had completed our family.

> "After he unwrapped my arm, he collected the sweat and measured the salt content. Dr. Shwachman then made the diagnosis – I had cystic fibrosis (CF)."

In 1974, Phyllis and I were accepted in a post-graduate program at the University of London - the first couple ever admitted. We lived for a year in Greenwich, near Greenwich Park, the Royal Naval Academy and the Royal Observatory. We had tutors in our disciplines. Phyllis became known in the U.K. as an expert in teaching dyslexic students and was the first non-British citizen to teach in a Dyslexic Center. I lectured on behalf of the American Embassy, at colleges and civic organizations, on the American Constitution and government. I met Archie Norman, M.D. at the Sick Children's Hospital, Great Ormond Street. Dr. Shwachman had recommended me to Dr. Norman. He

couldn't take me on as a patient but referred me to a private doctor who took adult CF patients. Phyllis and I and our two children were under the National Health Service. Jennifer was hospitalized for a urinary tract infection and I was admitted due to a major hemoptysis. We got excellent care at Greenwich Hospital and I from the CF doctor, all at no costs.

I took lectures on British politics at Nuffield College, Oxford University. Phyllis and I were awarded the title, "Associate, University of London."

We returned from our sabbatical year on the Queen Elizabeth II, since we were broke and Cunard accepted our American Express. I took an adjunct faculty position at Connecticut College and taught law on the graduate level there for thirteen years, before I retired from teaching in 1990. I had earned four graduate degrees, including a doctorate. Phyllis retired in 1992, with three graduate degrees. We traveled to the U.K. and Europe sixteen times, and to much of Canada, Alaska and Hawaii, where our daughter and her husband Eric live.

I lost the love of my life and best friend, Phyllis, to metastasized pancreatic cancer in August, 2009. Phyllis chose a "bucket list", rather than a second round of chemo and radiation as she had after a Whipple surgery two years earlier. We spent two weeks in Hawaii with Jennifer and Eric. Then Phyllis celebrated her seventy-fifth birthday at our favorite restaurant in London, Rules, with friends. Phyllis then had three more birthday parties, given by our cousins in Northampton, in Greenwich, where the neighborhood turned out for a dinner party, and at our flat, when her former tutor and his wife brought her a chocolate cake. Phyllis said to me shortly before she died, "Hal, seventy-five isn't a premature death." Such was her bravery. I have learned all about love in my eighty-one years, and as my favorite French song goes, to you I say, "I wish you love!" ▲

Hal is 81 and has CF. He is retired and continues to stay up on national and world events, from his home in Norwich, CT.

TILLMAN *continued from page 18*

in unlimited quantities in the laboratory, contains the delta-508 mutation, the gene responsible for about 70 percent of all CF cases and 90 percent of the ones in the United States. The tissue also contains the G551D mutation, a gene that is involved in about 2 percent of CF cases and the one cause of the disease for which there is now a drug. The technique could provide a powerful platform to screen drugs and study the biology of the disease.
http://tinyurl.com/83hypem

Computer-designed molecules point to new therapy for cystic fibrosis

By developing software that uses 3-D models of proteins involved in cystic fibrosis, a team of scientists at Duke University has identified several new molecules that may ease the symptoms of the disease. Computer algorithms created by the team predict how well a given molecular structure will block a basic protein-protein interaction known to occur in cystic fibrosis. To test the predictions, the scientists synthesized the molecules and measured how well they attached to one of the proteins in that interaction. The team then placed the best molecule into human cells with the cystic fibrosis mutation in a laboratory dish and found that their new drug blocked the protein-protein interaction and increased the cells' ability to balance salt and water levels. The results suggest that computers could make drug design for cystic fibrosis faster.
http://tinyurl.com/ceymtyn

Savara Pharmaceuticals' AeroVanc for cystic fibrosis patients achieves positive results in Phase I clinical trial.

Savara Pharmaceuticals, an emerging specialty pharmaceutical company developing innovative pulmonary drugs for the treatment of serious and life-threatening conditions, announced top-line data from its Phase Ia clinical study of the company's AeroVanc inhaled antibiotic. AeroVanc (vancomycin hydrochloride inhalation powder) is a novel dry powder form of vancomycin in a capsule-based inhaler for the treatment of respiratory methicillin-resistant Staphylococcus aureus (MRSA) infections in patients with cystic fibrosis (CF). By delivering vancomycin directly to the site of infection, AeroVanc is expected to improve clinical efficacy and reduce systemic exposure.
http://tinyurl.com/88avj2o

FDA Approves Pertzye

Digestive Care, Inc. (DCI), announced that the company has received U.S. Food and Drug Administration (FDA) approval of its New Drug Application (NDA) for Pertzye, indicated for the treatment of Exocrine Pancreatic Insufficiency (EPI) due to cystic fibrosis (CF) or other conditions. Pertzye is a unique pancreatic enzyme product containing bicarbonate-buffered enteric-coated microspheres. The Pertzye formula-

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WELLNESS

I Get Knocked Down

By Julie Desch

As you all know, life never is boring with CF. All of us eventually have to come to grips with an exacerbation. For some, it may only be every few years. Others go through it multiple times a year. One minute you are fine–maybe even great–and then BOOM, you feel like crap and it hurts to breathe. You're in the ER, someone is strapping O_2 tubing around your ears, they are searching for a vein to catheterize and, suddenly, you are not even remotely in control of your life. If you are like me, you might be wondering, "Where did this come from? What did I do wrong? Where is that pain medicine?" Maybe that last one is just me.

Like probably everyone reading this, I think the most infuriating aspect of this disease is the uncertainly. You can do everything right, and sometimes that just isn't enough. You get knocked down. Suddenly, the great groove you were in is moot. The fact that you were in week five of a 12-week workout program is irrelevant, as you now have a PICC and won't be lifting anything heavier than grocery bags for two weeks–if you are lucky. Your daily one-hour walks with your dogs and three-time weekly sprint interval sessions seem like ancient history as you are winded after a shower. There's not much to do about this. It is just what is.

The only choice available is in how to respond to the new state of your body. As I write this, I am pretty bummed (can you tell?). This is not going to be a sugar coated, pick yourself up, overly optimistic to the point of being irritating article. Instead, I am going to use my "Wellness" column to be very open about how I go through, 1) getting knocked down, and 2) getting up again. Maybe it's a good thing this happened because the article deadline is in eight days as I am writing this and, as of a week ago, I had no clue what to write about.

As you might guess by now, this was not a planned intervention. A week ago this time, I was lifting heavy weights and was experimenting with a treadmill interval program of rounds of hill sprints of 30 seconds, with 1 minute of walking recovery. My lungs were clear. I had just started my Cayston month, which always makes me happy. I was eating semi-Paleo and having great success by cutting all sugar and most grains and dairy. I was happy with my supplementation protocol. I was on a winning streak of over nine months off of IVs (this was a PR of several years.) My biggest stressor was my son's Spanish grade (oddly, this didn't seem to stress him a bit.)

So there I was, telling my partner, who was moving to a new home, "Don't lift that, you'll hurt your back. Let me do it!" I was moving boxes, clothes, and furniture–feeling amazing and strong. Then, there was this tiny headache and mild fatigue–neither surprising, given what we were doing. The fatigue turned into slight chills, so I took my temperature and sure enough, I had a low-grade fever. Huh? Weird. "Ok," I thought, "I'll just go easier tomorrow."

> "I think the most infuriating aspect of this disease is the uncertainly."

JULIE DESCH, MD

Well I certainly did! In fact, I was flat on my back in the hospital. In the middle of the night, I was in the ER with chest pain from Hell. Assuring the nurses that it was not cardiac, I nonetheless got the full throttle attention, including a chest x-ray, of course. Out of the blue, I had left lower lobe pneumonia. Triple antibiotics were hung, and I was thanking the Universe for the availability of Dilaudid.

My (foggy) thinking at this point was, "Oh well, this is no big deal. Here I go again on home IVs for a couple of weeks. I'll get through it as I have seemingly millions of times before."

The next morning, I was hit with Strike Two. One of the three antibiotics had sent my liver function enzymes

into the stratosphere. They were not just "elevated." They were frighteningly high. Two of the three drugs had to be stopped completely, because each was a potential suspect. I'll shorten the story here. The bottom line is that since I'm allergic to so many drugs I needed to wait until my liver recovered and then, essentially, take a guess at which of the two to retry, in order to know which drug to add to my (long) list of "don't even think about giving her this medication."

I will admit it. I became quite stressed at this point. As potential treatments for lung infections keep dropping out of contention, I got worried. I didn't really want to talk with family or friends at this point, because I knew I was on the verge of tears and I hate to cry about my health–it just seems so pointless. So instead, I broke down when my CF doctor called me (I was not at a CF center hospital at the time). Nice…

Eventually, the mystery was solved and IV aztreonam was declared a no-no for me. Strike Three. My darling Cayston is the inhaled form of aztreonam.

I was discharged on home IVs after five days in the hospital to begin the process of picking myself up and putting me back together. I'm starting to think of it as hardware "reboot." Here are my current steps in this process:

1) Create The Flowchart: There is so amazingly much to do when three IV antibiotics, each with a different time schedule, are added to the usual CF regimen. I know I'm preaching to the choir here, but when you are old, like me, it overwhelms. The only solution I've discovered is to use Tonic, an app I have previously written about, to schedule my (increased) treatments, IVs, supplements, feedings, exercise, sleep and everything else I need to do. With my iPhone telling me what to do when I need to do it, I can rest assured that I am not forgetting anything. This reduces a significant amount of stress.

If you don't have Tonic, the same thing can be done by simply creating a daily flowchart (this is what I used to do) and keeping it nearby to refer to.

2) Create Time: I know this is not physically possible. But, I really have to say NO to everything for a few days. It's never easy to do this, but the one thing I have learned over the years is that it pays to have nothing on the calendar for at least the first week home. Trust me, between follow up appointments, trips to the lab for copious blood draws and naps, this decision is justified very quickly.

3) Accept Help: I hate this one. Especially when it means having my kids stay with their other mom. But, with age comes wisdom (and sadly it seems that with age comes the need for more help.) I have learned that having the boys means grocery trips, errands and later nights than are optimal. Accepting help may also mean asking for others to bring food or shop. This may seem pathetic (it does to me.) But what is more pathetic, asking for help or letting pride interfere with health?

4) At least walk: Everyone is different in terms of how much exercise is best when recovering from an exacerbation. Some people can do box jumps outside their hospital room (these tend to be the young and very mobile people and may surprise the nursing staff a bit.) Others need complete absence of stress, including the "good" stress of exercise when recuperating. I fall somewhere in between. I need to move, not just for the purpose of expanding my fragile little lungs, but to maintain my sanity. I have a routine I follow when on IVs. As soon as the acute issue is dealt with and I am out of pain and can breathe, I walk every day. At first, this may be for only ten minutes at a time, several times a day. Over the course of the first week back, I build up to 20-30 minutes. It is slow and not pretty, but I am up and about, breathing fresh outdoor air. During the second week, I begin very simple lower body strength work, such as body squats and walking lunges. I also begin core work with planks and bridges. I do nothing with my arms while the PICC is in (I learned this the hard way.)

I truly believe it is essential to at least walk daily when you are on IVs, either at home or in the hospital, as soon as you possible can. Lying in a bed all day is the quickest way of losing strength. When you are sick and on IVs, your body is in a catabolic state, meaning muscle tissue is disappearing. The only way to offset this is to convince your body that you have no intention of not using that muscle, so it better not eat it for lunch. You do this by using the big muscles of your legs and trunk as soon as you can. This is very simply done by walking.

5) Plan Your Comeback: Maybe this is just me, but nothing lifts me out of my doldrums like planning my next workout scheme. I read about a system, or make up my own, and write out what I'm going to do each day. I've already discussed my activity action plan for the PICC days. I also come up with at least a month of exercise routines for the first month after I pull the PICC. What? Did she say she pulls her own PICC?

I also pick ONE good habit that I have let slip and work on improving that habit as part of my recovery. For instance, this time, I am reinstating

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TRANSPLANT TALK

If It Is Not One Thing, It Is Many

By Andrea Eisenman

There are days that I get so worked up about the minutiae that burdens my days, I forget how far I have come. In many ways it makes me feel "normal," like my peers and in other ways it makes me feel like I am taking things for granted.

I forget how lucky I am and how little my lungs bother me. Since my bilateral lung transplant, twelve years ago, not a day goes by that I do not silently thank my donor family and my doctors for saving my life. But not only saving my life, improving the quality of it immensely. It is just the rest of me that is falling apart.

Let me illustrate by writing about my last six months and the domino effect of having one procedure that seemingly led to more problems and none anticipated. I had a birthmark removed, after much debate from my dermatological surgeon. She had just recently removed two large basal cell carcinomas from my scalp, twelve and six months prior, by way of Mohs surgery. Since being on immuno-suppressants, I have had many squamous and basal cell cancers removed. The birthmark in question was on my right leg, under my knee. It didn't hurt or look that different but due to its size and its edges becoming irregular, the surgeon felt it should come out so it wouldn't become cancer. She made it seem that it would be "no big deal" to remove it. It was a minor cut and one stitch. Nothing could have been further from the truth. She told me it would be nothing like having Mohs surgery. It was worse than having Mohs surgery!

Usually, with Mohs surgery, I would not be allowed to walk very far, three blocks per day and I would need to rest so as not to send too much blood to the area of the incision. Pumping too much blood to a newly stitched area can cause swelling and then scarring later on. On one's scalp, that can mean a bald spot. So, when I had these two areas removed, it was very difficult for me not to do my

ANDREA EISENMAN

usual exercise and walking or taking the stairs in the subway. But it is only for two weeks while the stitches are in and one week after stitches come out. When I had the birthmark removed, it was much more painful than I had expected. I had to keep my leg up for several days and when I was able to walk, I limped around. Since the stitches were on my leg and I walk on my leg, they were not able to heal as much as something on someone's head or arm. I went to get them removed after two weeks to find that they had to stay in an additional week, but that was only after they started to remove some of them and the wound popped open. Still limping for another week and not allowed to exercise, I had the stitches removed and the wound immediately opened, once the bandage came off.

During all of this, I had started to get severe pain in my hip, specifically the right one. I thought it was inflammation or arthritis. It started to hurt so much, I was icing it constantly. Finally, after about six weeks of pain, difficulty in walking or playing tennis and especially when walking up stairs, I went to see an orthopedist for my hip. I even had a hard time sleeping because, unless I slept on my back, my hip hurt. I had to buy a foam bed-topper just to get some sleep without waking up in pain every hour.

I finally saw the orthopedist who, after looking at my x-rays and poking me in my hip, diagnosed me with hip bursitis. He gave me shot of cortisone—not pleasant. But afterwards, it felt wonderful. With cortisone, I had high blood sugars for about four days due to my post-transplant diabetes. It was a small price to pay to be pain-free. I was to start physical therapy (PT) for about two months, twice a week.

Before starting PT, I met with a physical therapist. We talked about what might have led up to the hip bursitis and I thought it could have been the removal of the birthmark. Since I had been limping around for so many days and walking funny, I felt it threw my whole body off. She agreed that it probably started it off. She also could see that my right leg was not as strong as my left leg.

Currently I am almost done with PT but when I started, I had to explain my back situation to the physical therapist. When I was 16, I had a

spinal fusion for severe scoliosis. I had been to PT for my back two years prior and they were not really able to help me much. So, I was dubious about my hip being fixed. I went diligently twice a week to PT and did my home exercises as prescribed. After a few sessions, my back went out as it had not done before in my memory. Then after going to PT it happened again. This time, I could barely walk and had to take muscle relaxants for five solid days. The physical therapist, not used to someone with such limited range in their back was stretching me in a way that hurt my nerves in my back. Once we discovered what was going on, I told her that we cannot do those stretches for my IT band and then the PT was able to work. It had now been about three months since I started PT and I am feeling much better in my hip, pain is much diminished and my legs feel stronger.

Around the same time I started PT, I got a sty in my eye. I had no idea what it was, but it felt huge and a fever accompanied it. Of course, it blew up on a Friday night so instead of festering in city ER, I went to a walk-in clinic. Can you imagine what it is like to have to explain all my issues—lung transplant, CF, diabetes and medications to people used to treating maladies like a cold or bronchitis? They told me to see an eye doctor but gave me antibiotic drops and to take Tylenol for my fever. I then went to see an eye doctor in Long Island, near my mom whom I was visiting. I had never met him before and he checked me over and told me he would suggest I let him lance my cyst to drain the pus and allow it to heal. Now, those of you who do not know me don't know what a wuss I am about my eyes and I really debated running out of his office and never coming back. But a calm came over me, plus wanting this thing healed as soon as possible, and I told him he could lance it. He pulled out a blade and had to slice twice. Meanwhile, I am not supposed to move, for obvious reasons. Then, he had to squeeze the pus out which was the most intense pain, even more than the hip, I had in a while. I was to go home, use a warm compress twice a day but more if I had the time and then put two kinds of drops in my eyes for two weeks. One was prednisone and one was an antibiotic. Plus, since my eyes were dry, I had to also put the fake tears in. I felt traumatized by this ordeal.

Between doing my PT exercises and using eye drops and putting warm compresses on my eye, I was a mess. I still had all my usual inhalation, nasal lavage stuff as well. I was good about all of the eye care. But for some reason, after a month, the sty came back. I decided to see my regular eye doctor at this point, since I knew it was not an emergency. He told me to continue with fake tear drops and warm compresses for a while longer. So far, so good.

Even though, these are all non-life threatening things, they piled up and made me frustrated. It is great not to have to worry about whether I can breathe, or whether I will have to start IVs for an infection, I am grateful. But the small things can become big things when they all happen at once. And since pain was involved, it distorted my usual sense of, "everything will be ok." For the future, I will try to remember how far I have come - dealing with my CF and life post-transplant. I really should be able to handle everything at this point. ▲

Andrea is 47 and has CF. She is a Director of USACFA and is the Executive Editor of CF Roundtable and Webmaster. Her contact information is on page 2.

CLUB CF ONLINE

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

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

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

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

COUGHING WITH A SMILE...

Chatty Chatty Bang Bang

By Jennifer Hale

Hello readers of *CF Roundtable*. Hope this issue finds ya'all doing and feeling well! The topic of this issue is about what you do while you are doing your treatments and what has worked. Well, for me, I simply watch TV or surf the net so writing a whole column on that was not going to take up much space- LOL. So I thought I would write about CF forums and chat rooms and what I personally thought of them. Again, this is solely my opinion and I can always stand corrected. It's all good!

Growing up I did not want people to know I had CF. I was luckily not real sick growing up and it was easy to avoid talking about CF, and I sure didn't look like I was sick. I still don't look like I have CF, but I am much worse as an adult health-wise. Funny you can never tell a book by its cover! I guess that is a whole other story. (Maybe I found my next column topic.) Anyhow, I was not into sharing my disease, promoting awareness about it or even talking to others with CF. I did not go to the camps for CF patients. (Now I am dating myself! But that is good when you have CF, yeah 40!) I just did not want CF to be so much a part of my life, including being around or even talking to others who had it. I was not encouraged to do so by my parents and therefore I just did not find a need to have that connection and/or interaction.

When I was growing up, kids with CF did not fare as well as kids do now. There weren't all the wonderful medications and treatments like there are now and those with CF had a very uncertain future. We still have an uncertain future and I don't want to make it appear that having CF is a walk in the park, but the meds and therapies that are out in the year 2012 are *a lot* more advanced than in 1972.

I remember one time I was in the hospital for an exacerbation and a friend was doing a report and she chose to write about CF. I was mortified and nervous that EVERYONE at school would know why I was in the hospital and then they would know that I had the disease she was reporting on for her school paper. I was very upset and, of course, when I got back to school no one even batted an eye. That was how secretive I was about my disease. I just did not want anyone to know. I suppose I just did not want to be perceived as different from everyone else. That was my choice on how to live my CF teen years but that all changed as I got older.

> *"As I got older my health has declined, but my need to talk about my CF and and to share ideas with other people with CF has increased."*

JENNIFER HALE

As I got older my health has declined, but my need to talk about my CF and make people aware of what I go through and to share ideas with other people with CF has increased. I found I enjoy talking to people about my CF experience and even more I enjoy talking with others who have CF or whose loved ones have CF. Being 40 with CF, I have been around the block and to give hope to families whose small children have CF has been one of my greatest accomplishments. I now love talking about CF with people and sharing my story and experiences. It is a far cry from what I used to be like growing up, but this hope that I can share with others is sometimes just what that other person needs and that is a great feeling. This leads me to my opinion on forums and chat rooms for CF.

There are a couple of these chat rooms and forums to peruse in the CF community and I have actually met some really great people who I now talk to not only on the forum but also via phone. It has been great to share our stories, experiences and even

laugh at some of the things CF people go through. It is really comforting to talk to someone who "gets it" and who has been through the same things I have, as well as things I haven't experienced. I get their honest opinion and first-hand experience of a medication, procedure or therapy and that is priceless information. Yes, the doc can explain things thoroughly but the doc has not gone through it and "felt" it, so it is nice to ask my friends in the CF community what the real deal is.

Forums and chat rooms are GREAT to find some good information but I also feel one has to precede with caution. There are a lot of people on the boards with good ideas *and* some with really strange ones. Some people are really desperate in their CF walk and I personally feel desperate people sometimes make desperate choices. I feel you need to ask your doctor about any treatment or therapy before adding it to your routine.

There are some crazy ideas out there and people putting some crazy solutions in their nebs. But that is just one example. I also feel you can get wrapped up in someone's life a little too much which could add much stress to your own. So again, when I go on these forums I really try to listen a lot and proceed with caution. I remember one time I got so wrapped up in this one person's CF drama. A whole bunch of people on the forum were praying for this person, including myself. Come to find out the person was lying about having CF and it was this whole big story. I felt horrible. I mean I was praying for this person and looking at the updates daily and then come to find out the person was not afflicted with CF. WOW! Like I said, red light, green light, YELLOW LIGHT!!!!!

On a better note, like I said before, I have met some great people through the forums and formed some real solid relationships. I have learned a lot from these forums and have asked my doc a lot of questions over the years just from information I read on the forums and chat rooms. Since we cannot be physically around someone with CF, having these places to go is a great alternative. It is a place where you can ask any question about what you're dealing with and you will get all kinds of responses. Some good, some bad and some that make you really think. It is a good resource to have at your fingertips and that is exactly how I use it – as a resource. I love that some people have emerged to become great friends and that is a bonus! The days of CF camps are over so now we have virtual camps and for the teens out there that is really a great thing. I know some people out there don't have a lot of support in their CF journey and these forums can provide support for them.

All in all, I am glad I am more open talking about my CF and it is nice to meet others from literally all over the world who are also dealing with the same issues I am on their CF journey. Hopefully we will all be able to share and support each other through the good times, bad times and eventual cure for CF! ▲

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

Information For People Who Travel On Airlines

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

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

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

IN THE SPOTLIGHT

With Jordan Herskowitz

By Jeanie Hanley and Andrea Eisenman

I had the pleasure of meeting our newest interviewee two years ago when he first moved to the New York area. We had lunch in Chelsea over dumplings and soup. He told me about his life with his two siblings who have CF. And I wondered how it would be to live as the "healthy one" amongst two brothers who had a fatal disease. How might that dynamic shape someone? He struck me as a funny, bright and talented young man with a lot of drive. He told me about his show, *Growing Up Jordy Pordy*, and encouraged me to come see the production in a local theatre. I was glad I went. I laughed and cried, and cried some more. The show was very moving, not only for me but for my friends whom I brought along. Seeing Jordan morph into numerous characters on stage, I felt it helped explain some of the highs and lows of living with CF as seen through a non-CF sibling and extended family members. After seeing his show, I could see that Jordan would have a bright future in theatre. Please welcome our latest star, spotlight please!

Name: Jordan Herskowitz (Jordy Pordy)
Age: 25

Occupation: Creative Director, *Jordy Pordy Productions*, Executive Director, *Richie's Spirit Foundation*

Religious or spiritual?

I have a strong cultural connection to Judaism, but I'm not terribly religious observant. I love learning about other religions and consider myself a spiritual person.

Where did you grow up? Where do you live now?

JORDAN HERSKOWITZ

I was born and raised in Dallas, Texas. I went up north for college (to Oklahoma!), and finally made my dream of living in New York City real two years ago.

What was in like growing up with two siblings with CF?

Because hospitals and doctor visits came to be so routine for my family, I never realized how different my family was from others when I was younger. My parents did a great job making sure we stayed humble and we're grateful for what we had.

You were also the middle child, did you feel invisible?

I definitely yearned for attention, and started finding other outlets as a kid to get that attention. I became the class clown in school and was always putting on a show for my family or friends, whenever I had the chance.

At what age were your siblings diagnosed?

My older brother, Neil, was diagnosed at six months and Richie, my younger brother, was diagnosed at birth.

How is the health of your brothers?

My younger brother, Richie, became the youngest CF patient to receive a bilateral lung transplant in 1996. He lived almost a decade longer than doctors expected but passed away in October 2007. My older brother, Neil, is 26 years old and is doing great. He has a much more mild case than Richie did and is on medicines and nightly breathing treatments that help keep him healthy.

How did your brothers' health affect you?

Neil and Richie have never let their CF define who they are. It has been their strength, courage, and positive outlook that has made me truly treasure life and the blessings each one of us has, instead of what we don't have.

Do you think your brothers' CF changed you and if so, how?

Absolutely. Growing up with brothers who have a terminal illness definitely adds to the family dynamic. I believe I've become a more independent, compassionate and caring person because of what my family has been through.

What is your show about?

Growing Up Jordy Pordy tells the story about my life as a professional

mascot. But once my furry gloves come off, I reveal my own struggles of growing up Jewish in Texas, being the forgotten middle child with both of my brothers born with cystic fibrosis, and my family's journey with organ donation. *Growing Up Jordy Pordy* defies the lines of theatre norm, as I play over 30 characters.

Do people have to know about CF to enjoy *Growing Up Jordy Pordy*?

Not at all! The great thing about the show is its universal appeal. Audiences find something unique to connect to in the show, and also something new they didn't know about before.

Richie's Spirit Foundation, how did it come about? Where do funds go?

When I started touring with my show, many people took interest in organ donation and helping to keep Richie's spirit alive. And so, I decided to start Richie's Spirit Foundation, which promotes organ donation and shares the positive outlook Richie had with others. In three short years, the foundation has grown from a small start-up to a nationally recognized organization. Our grants include a College Scholarship and a Transplant Assistance Fund and events include a basketball tournament and my one-man show.

How did you get the nickname?

Grammie, my spunky, 85-year-old grandmother, gave me the nickname. She has a nickname for each of her nine grandchildren. There's no great story behind it except that Grammie thought it rhymed well.

Did you always want to be a performer?

I've always had a fascination with performers and the entertainment world. Especially in today's busy world filled with technology, I believe theater is one of the great ways of keeping human interaction and storytelling alive and thriving.

How did you get into doing your show?

I graduated from The University of Tulsa with a degree in Theatre in 2009. During my junior year, I received a grant to write my show over the summer. What was meant to be just a senior project turned into something much bigger!

How long have you been doing the show?

I've done over 300 performances of the show around the United States, United Kingdom, and South Africa.

What do you hope people take away from it?

I hope people take away an understanding of cystic fibrosis and the importance of organ donation. I've always said that after all of the shows I've done, if one person decides to become an organ donor and saves someone's life (like my brother's,) it was all worth it.

Tell me about your workshops?

Besides acting and writing, I love teaching and sharing my talents with students and adults of all ages. I lead basic acting workshops, grief-based drama workshops, and personal narrative writing workshops. I've led these around the country and in England.

What is your inspiration?

I'm constantly being inspired by people I meet, things I see/read, performances I watch. For my show, I was inspired by Billy Crystal, whose one-man show I saw in New York City when I was in high school.

If you could change anything on your life, what would that be?

Though there have been many challenges I've had to face, I wouldn't change a thing in my life! I believe those challenges have made me who I am today.

Do you think there will be a cure for CF in your life-time?

It's amazing the progress CF research has made in the past 20 years, and I'm keeping hope up that a cure will be found in my lifetime.

Where do you see yourself in five years?

I am working on a new one-man show and would like to go to grad school, but it's hard to plan the future!

Where have you performed?

I've performed at the CFRI conference and retreat, Donate Life conference, the Edinburgh Fringe Festival in Scotland, and many other venues and festivals. For more information about the show and my work, visit www.jordypordy.com.
For more information on Richie's Spirit Foundation, visit www.richiesspirit.org.

Jeanie Hanley is 50 and is a physician who has CF. She is a Director of USACFA. Her contact information is on page 2. Andrea Eisenman is 47 and has CF. She is a Director of USACFA and is the Executive Editor of CF Roundtable and Webmaster. Her contact information is on page 2.

If you would like to be interviewed for this column, please contact either Jeanie or Andrea. ▲

DESCH *continued from page 27*

my policy of morning meditation, which I have let slide over the last few months. In some way, this makes the illness an initiator of positive change. This takes some of the "helpless" feeling of getting sick for no apparent reason away. At least I can use it for a good purpose.

6) Sleep More Than You Think Is Necessary: This doesn't really need much explanation. Healing bodies need sleep. It is during sleep that your cells package up and send out the trash. A CF exacerbation leads to a lot of cellular trash. It is like spring-cleaning and sleep is essential. Sleep also helps the immune system do its thing. Nine hours at night is the minimum for me. An afternoon nap is a daily bonus point.

7) Stay In Contact: Isolation is a quick way into depression and increasing stress hormones. This acts as a break on the healing process. While you may not look or feel like interacting with others, it is not a good idea to hole up. I am the worst example of this and I'm actively trying to reach out to others this time around. Writing Facebook posts doesn't count. Actual human encounters, in person or over the phone or Skype, lead to a feeling of connection and it is this connection that has proven invaluable in positive emotional health.

8) Remember What Works: Finally, while there may be many things that are functioning less than ideally (thank you lungs and liver) I try to remember each day what is still working pretty well, even after 51 years playing the game of life with CF. Each day, I write down at least three things that are going well. Today, the coffee tasted fabulous (thank you taste buds), I was able to do the full Tai Chi workout of 35 minutes this morning and I had a great night of sleep last night.

So yes, I do get knocked down. Sometimes, it seems as though it is by a flying railroad tie that hit me from behind, while I was innocently chasing butterflies in a field of lilies under a cloudless sky. But, my son put it very nicely when he said, "Don't worry Mom, you'll get up again. You always do." ▲

Julie is 51 and is a physician who has CF. She may be contacted at: jdesch@usacfa.org.

TILLMAN *continued from page 25*

tion was previously marketed by DCI for over a decade under the trade name Pancrecarb MS-16.
http://tinyurl.com/6o9jn4y

Cystic fibrosis lung infection drug trial gets OK'd after FDA lifts clinical hold

The U.S. Food and Drug Administration agreed to lift a clinical hold on a mid- to late-stage drug to treat cystic fibrosis patients for two different types of lung infections nine months after invoking it. Arikace is designed to treat cystic fibrosis patients who have lung infections from nontuberculous mycobacteria (NTM) and those with chronic pseudomonas aeruginosa lung infections. Insmed believes the drug can fill a growing unmet need for cystic fibrosis patients by delivering high, sustained levels of amikacin directly to the lung, potentially providing sustained improvement in lung function and improvement in patient symptoms with a once-a-day dosing regimen. If approved, the drug could be the first inhaled antibiotic for cystic fibrosis patients for once-daily administration. Arikace has received orphan drug designation for pseudomonas aeruginosa patients, but Insmed is awaiting one for NTM patients.
http://tinyurl.com/7c82s7m

Cystic fibrosis breakthrough reveals why females fare worse than males

Researchers have published a study which represents a major breakthrough in understanding why females with cystic fibrosis do worse than males. The study is the first to show that the female hormone estrogen promotes the presence of a particular form of bacteria which results in more severe symptoms for female cystic fibrosis patients. In addition, females who were taking the oral contraceptive pill, which decreases the amount of naturally occurring estrogen in their bodies, were found to have lower levels of the problematic bacteria. The research found that estrogen promoted the presence of a 'mucoid' form of the bacterium Pseudomonas aeruginosa in the lungs of women with cystic fibrosis.
http://tinyurl.com/79o6ohv

Garlic Constituent Blocks Biofilm Formation, Could Benefit Cystic Fibrosis

Tim Holm Jakobsen and Michael Givskov of the University of Copenhagen, and their many collaborators have pinpointed a constituent of garlic that attacks a key step in the development of biofilms, in an effort they hope may offer help in particular for patients with cystic fibrosis. Ajoene, the major constituent of a multitude of sulfur-containing com-

TILLMAN *continued from page 34*

pounds produced when garlic is crushed inhibits expression of 11 genes that are controlled by quorum sensing in P. Aeruginosa. Ajoene also reduces the production of rhamnolipid, a compound that shields the biofilm bacteria from the white blood cells that otherwise would destroy bacteria. By combining ajoene with the antibiotic tobramycin, it's possible to kill over 90 percent of bacteria living in a biofilm. Garlic not only has antibacterial properties; it has anti-viral, anti-fungal, and anti-protozoal properties as well, and it has beneficial effects on the cardiovascular and immune systems.
http://tinyurl.com/bt4ufky

Top-Line Data from Phase 3 Trial of Ataluren in Patients with Nonsense Mutation Cystic Fibrosis Show Promising Results

PTC Therapeutics, Inc. announced the results from a Phase 3 study of ataluren, an investigational new drug, in patients with nonsense mutation cystic fibrosis (nmCF). The results demonstrated positive trends in the primary endpoint, lung function as measured by FEV_1 (forced expiratory volume in one second), and in the secondary outcome measure, rate of pulmonary exacerbations. Results also showed a substantial ataluren treatment effect in FEV1 and exacerbation rate in the pre-specified subpopulation of patients not receiving chronic inhaled antibiotics. Ataluren is a protein restoration therapy designed to enable the formation of a functioning protein in patients with genetic disorders caused by a nonsense mutation. A nonsense mutation is an alteration in the genetic code that prematurely halts the synthesis of an essential protein. The resulting disorder is determined by which protein cannot be expressed in its entirety and is no longer functional, such as the cystic fibrosis transmembrane conductance regulator protein (CFTR) in nmCF.
http://tinyurl.com/7a2zuwb

Bronchitol Reduces Risk of Exacerbations in Patients with Cystic Fibrosis

Pharmaxis presented a new analysis of its two Phase 3 clinical studies of Bronchitol® showing positive trends in reducing exacerbations in all age sub groups of cystic fibrosis patients. Bronchitol has been developed to help clear mucus, improve lung function and reduce exacerbations in patients with cystic fibrosis. Bronchitol is a proprietary formulation of mannitol administered as a dry powder in a convenient hand-held inhaler. Bronchitol hydrates the lungs, helps restore normal lung clearance, and allows patients to clear mucus more effectively. Clinical studies have shown Bronchitol to be safe, effective, and well tolerated.
http://tinyurl.com/6ur75hz

Continued on page 36

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.

LUNG TRANSPLANT

Duration of Voriconazole Exposure: An Independent Risk Factor for Skin Cancer After Lung Transplantation. Fiona O. Zwald MD, Margaret Spratt MD, Bianca D. Lemos MD, Emir Veledar PhD, Clint Lawrence MD, George Marshall Lyon MD, Suephy C. Chen MD, MS. Dermatologic Surgery. Article first published online: 2 MAY 2012

Duration of voriconazole exposure correlates with number of nonmelanoma skin cancers (NMSC) after lung transplantation. All patients exposed to voriconazole should be educated about their increased risk of skin cancer and should have regular dermatologic follow–up for skin cancer screening. Physicians caring for lung–transplant recipients should consider alternatives to voriconazole in patients at risk for skin cancer.
http://tinyurl.com/c2lf28q

Antibiotic Improves Function After Lung Transplantation

Azithromycin, a broad-spectrum antibiotic with anti-inflammatory properties, may be an effective treatment for bronchiolitis obliterans syndrome (BOS), a life-threatening complication that occurs after lung transplantation.
http://tinyurl.com/7uyfkhs

Diabetes mellitus and survival in cystic fibrosis patients after lung transplantation. Markus Hofer, Christoph Schmid, Christian Benden, Rudolf Speich, Ilhan Inci, Walter Weder, Annette Boehler. Journal of Cystic Fibrosis. Volume 11, Issue 2, Pages 131-136, March 2012

One– and five–year survival rates after lung transplantation tend to be better in CF recipients with Diabetes mellitus compared to those without Diabetes mellitus. No impact of Diabetes mellitus on the development of Bronchiolitis obliterans syndrome was found.
http://tinyurl.com/7jxtnco

Sinonasal persistence of Pseudomonas aeruginosa after lung transplantation. J.G. Mainz, J. Hentschel, C. Schien, N. Cramer, W. Pfister, J.F. Beck, B. Tümmler. Journal of Cystic Fibrosis. Volume 11, Issue 2, Pages 158-161

The report demonstrates persistence of identical P. aeruginosa genotypes in CF upper airways prior to and after lung transplantation underlining risks of descending colonization of transplanted lungs with P. aeruginosa, which increases risks of graft dysfunction. Therefore, the authors recommend regular assessment of sinonasal colonization prior to and after lung transplantation. Sinonasal inhalation with antimicrobials should be investigated in prospective trials.

http://tinyurl.com/88zlspu

TREATMENTS

Long-term effect of chronic intravenous and inhaled nephrotoxic antibiotic treatment on the renal function of patients with cystic fibrosis. Marius C. Florescu, Elizabeth Lyden, Peter J. Murphy, Diana F. Florescu, Jennifer Fillaus. Hemodialysis International. Article first published online: 3 APR 2012

Chronic inhaled colistin and gentamicin are associated with an increase in blood urea nitrogen but not creatinine at the end of follow up. Inhaled colistin was associated with episodes of acute kidney injury. Well–managed intravenous use of nephrotoxic antibiotics in cystic fibrosis population is associated with no major long–term renal toxicity.
http://tinyurl.com/83ue73p

Ivacaftor potentiation of multiple CFTR channels with gating mutations. Haihui Yu, Bill Burton, Chien-Jung Huang, Jennings Worley, Dong Cao, James P. Johnson Jr., Art Urrutia, John Joubran, Sheila Seepersaud, Katherine Sussky, Beth J. Hoffman, Fredrick Van Goor. Journal of Cystic

Fibrosis. Volume 11, Issue 3, Pages 237-245, May 2012

The in vitro data suggest that ivacaftor has a similar effect on all CFTR forms with gating defects and support investigation of the potential clinical benefit of ivacaftor in CF patients who have CFTR gating mutations beyond G551D (G178R, S549N, S549R, G551S, G970R, G1244E, S1251N, S1255P and G1349D).
http://tinyurl.com/czwudxd

Inhaled aztreonam lysine for cystic fibrosis. Lindsay E Hoffman and Denise K Lowe. Journal of Pharmacy Technology. 2012;28:75-81

Aztreonam lysine is a newer inhaled antibiotic that improves forced expiratory volume in 1 second, respiratory symptoms, and quality of life measures when used as maintenance therapy in patients with cystic fibrosis (CF) and chronic P. aeruginosa infection.
http://tinyurl.com/7jthwqk

Effect of azithromycin on systemic markers of inflammation in cystic fibrosis patients uninfected with Pseudomonas aeruginosa. Felix Ratjen, Lisa Saiman, Nicole Mayer-Hamblett, Larry C. Lands, Margaret Kloster, Valeria Thompson, Peggy Emmett, Bruce Marshall, Frank Accurso, Scott Sagel and Michael Anstead. Chest. Published online before print May 17, 2012

In patients not infected with P. aeruginosa, oral azithromycin significantly reduced neutrophil counts and serum inflammatory markers within 28 days of initiating treatment.
http://tinyurl.com/7zzpm8k

Effect of dornase alfa on inflammation and lung function: Potential role in the early treatment of cystic fibrosis. Michael W. Konstan, Felix Ratjen. Journal of Cystic Fibrosis. Volume 11, Issue 2, Pages 78-83, March 2012

A recent analysis found that initiation of dornase alfa has the potential to alter the course of cystic fibrosis by decreasing the rate of lung function decline in children and adults. These encouraging results, possibly linked to indirect effects on inflammation, suggest a greater role for dornase alfa therapy in the early treatment of CF, where it may help preserve lung function and potentially extend survival.
http://tinyurl.com/7k97vlj

Clarithromycin therapy for patients with Cystic Fibrosis: A randomized controlled trial. P. Robinson MB BS, MD PhD, Michael S. Schechter MD, MPH, Peter D. Sly MB BS, MD DSc, Kaye Winfield, Julie Smith, Siobhain Brennan, Masaharu Shinkai MD, PhD, Markus O. Henke MD, Bruce K. Rubin MEngr, MD, MBA. Pediatric Pulmonology. Volume 47, Issue 6, pages 551–557, June 2012

The authors conclude that clarithromycin is not effective in treating cystic fibrosis (CF) lung disease.
http://tinyurl.com/7remr5w

Azithromycin maintenance therapy in patients with cystic fibrosis: A dose advice based on a review of pharmacokinetics, efficacy, and side effects. Erik B. Wilms PharmD, Daniel J. Touw PhD, PharmD, Harry G.M. Heijerman PhD, MD, Cornelis K. van der Ent PhD, MD Pediatric Pulmonology. Article first published online: 3 JAN 2012

Due to the extended half–life in patients with cystic fibrosis (CF), the weekly dose of azithromycin can be divided in one to seven dosing moments, depending on patient preference and gastro–intestinal tolerance. No important side effects or interactions with other CF–related drugs have been documented so far.
http://tinyurl.com/7auy8yd

Continued on page 38

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.**

Successful treatment of cepacia syndrome with a combination of intravenous cyclosporin, antibiotics and oral corticosteroids. Francis J. Gilchrist, A. Kevin Webb, Rowland J. Bright-Thomas, Andrew M. Jones. Journal of Cystic Fibrosis. Published online: 03 May 2012

Burkholderia cepacia complex (BCC) is a group of 17 closely related bacterial species that can cause pulmonary infection in patients with cystic fibrosis (CF). The clinical manifestations of BCC infection are varied but can include cepacia syndrome, which is a rapidly progressing necrotising pneumonia with an almost universally fatal outcome. Aggressive treatment with a combination of 4 intravenous antibiotics, oral corticosteroids and cyclosporin brought about clinical, radiological and biochemical resolution of one patient's cepacia syndrome. This case highlights the possible role of cyclosporin in the treatment of cepacia syndrome.

http://tinyurl.com/7ucmhqs

Inhaled Aztreonam Lysine for Cystic Fibrosis. Lindsay E. Hoffman and Denise K. Lowe. Journal of Pharmacy Technology. 2012;28:75-81

Aztreonam lysine is a newer inhaled antibiotic that improves forced expiratory volume in 1 second, respiratory symptoms, and quality of life measures when used as maintenance therapy in patients with CF and chronic P. aeruginosa infection. Availability of aztreonam lysine only from specialty pharmacies may limit its use.
http://tinyurl.com/7jthwqk

SINUS

CT of the paranasal sinuses is not a valid indicator for sinus surgery in CF patients. Jacob Rasmussen, Kasper Aan, Rikke Norling, Kim G. Nielsen, Helle Krogh Johansen, Christian von Buchwald. Journal of Cystic Fibrosis. Volume 11, Issue 2, Pages 93-99, March 2012

There was no significant correlation between the CT score and detection of pus, pathogenic bacteria or symptoms. Pus and pathogenic bacteria were found in several cases without sinus opacification on the CT scan. Non pathogenic and sterile cultures were also found in sinuses with opacification. A CT scan is not a valid criterion for sinus surgery in cystic fibrosis (CF) patients.
http://tinyurl.com/7gpvmzr

FYI

Canadian Airway Clearance Study of Positive Expiratory Pressure (PEP) Mask vs. High-Frequency Chest Wall Oscillation (HFCWO)

The Canadian national study suggests that the PEP mask is superior to HFCWO as a mode of physiotherapy in patients with CF.
http://tinyurl.com/6s59p5w ▲

Laura Tillman is 64 and has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.

In Memory

Leslie Bryant Alford, 51
Baytown, TX
Died June 12, 2012

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.
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KATHY RUSSELL 5/13
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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.

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- Please let us know of the major occurrences in your life (*e.g., marriages, births, completion of educational degrees or training, career advancement, transplants, anniversaries, birthdays*) and we will print your information in **Milestones**.
- Share your ideas for **Focus Topics**, feature articles or any suggestions for improvements you may have to help make *CF Roundtable* more relevant and interesting to you.
- You can reach **USACFA** and ***CF Roundtable*** at anytime by phone **(248) 349-4553**. You may email us at **cfroundtable@usacfa.org**
- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq., 712 Main, Suite 2130, Houston, Texas 77005. Email: cflegal@cff.org**.
- You may subscribe at cfroundtable@usacfa.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.

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

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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

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