CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
WINTER 2013

Bone Health In People With CF

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Bone health has largely been investigated in the elderly and post-menopausal women because of the increased fracture risk in this population. In more recent years, the diagnostic tools and treatments for bone disease in this population have been extended to children, adolescents, and younger adults and to populations with chronic diseases. The application of these tools and treatments to younger populations, including those individuals with CF, continues to evolve.

Bone is composed of bone cells, collagen, calcium and phosphorus. Bone accrual occurs steadily through childhood, and 40-50% of bone mineral accrual occurs during the teenage years into the early twenties. During this process bone is built, bone is broken down, and bone is rebuilt to improve the strength of bone. The majority of this bone accrual is determined by genetics, but factors such as diet and exercise contribute. With aging, the amount of bone that is broken down exceeds the amount of bone that is built, and individuals whose bone accrual during the teenage years is sub-optimal are at increased risk for osteoporosis later in life.

> "In at least some individuals with CF, optimal bone density is not achieved during puberty and the bone loss of aging occurs earlier."

Dual X-ray absorptiometry (DXA) was developed to screen for compromised bone health in the elderly. DXA uses low dose radiation to measure bone density in the spine, hip, and whole body. DXA measures "density" of bone but does not indicate the quality of bone. DXA results are presented as standard deviation scores. The term BMD-Z is used for children, men under the age of 50, and pre-menopausal women while BMD-T is used for older men and post-menopausal women; (a BMD-Z/T of 0 is the 50% and of -2 is the 2.3%.) Interpretation of BMD-Z in children can be challenging. In order to arrive at a Z-score children are compared to children of the same sex and

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CF ROUNDTABLE
FOUNDED 1990
Vol. XXIII, No. 1

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EDITOR'S NOTES

Fortunately, winter has been none too bad here in Oregon. I have done my best to stay away from large groups of people where I might be exposed to FLU. So far so good. I hope that you have been as fortunate.

We have sad news to share. After a valiant battle against infection, **Debbie Ajini** died on October 26, 2012. She had just written of her transplant and how full of hope she was. See her obituary on page 20. Our sympathy goes out to all of her family and friends. We will miss her.

I hope you already have read **Dr. Andrea Kelley's** article that starts on page 1. She discusses bone loss and ways to prevent it. Our Focus topic: "Osteoporosis, Osteopenia And Other Skeletal Issues" continues with an article by **Anne Williman.** She tells of taking various meds to deal with bone loss. **Nicole Matthews** tells of her experience with bone pain and how she tries to avoid it. **Kathy Russell** adds her observations on bones and aches in "Speeding Past 50".

In "Ask The Attorney", **Beth Sufian** answers questions from our readers regarding Social Security benefits. **Julie Desch** uses her "Wellness" column to address aging with CF. She uses the term: "Senior Cystic" to describe herself. In "Spirit Medicine", **Isabel Stenzel Byrnes** writes of how being honest is healthy for us. As always, **Laura Tillman's** compilation of "Information from the Internet" is chock full of news.

"Coughing With A Smile" finds **Jennifer Hale** explaining Stand Up Paddle Boarding (SUP). Sounds like fun. "In The Spotlight" features **Stephanie Rath**. (At the time of the interview, Stephanie had not yet been elected to the Board of Directors of USACFA. We are pleased that she works with us now.)

We have three articles in "Voices from the Roundtable" in this issue. **Brandi Edelman** writes of expecting to be treated with the respect that is due us. **Chris Kvam** discusses living with CF rather than fighting CF. **Steve Jenkins** writes of his dissatisfaction with some portions of the organ transplant policies.

Be sure to check out page 17 for the latest offering from **Breathing Room** then turn the page to see pictures in "Our Family Photo Album". When you look at the "Milestones" on page 10, notice that people who have CF are living longer and doing wonderful things with their lives. Then take a look at the letters in the "Mailbox", I think you will find some of them to be very interesting.

We'd like to take this opportunity to thank all of our donors and especially our benefactors who can be found on pages 5 and 11. We couldn't do this without you.

Be sure to look at the upcoming Focus topics, in "Looking Ahead", on page 3. Perhaps you have some experience you'd like to share with the readers. Please write to us and let our readers hear from you.

Here's to a lovely and pleasant spring!
Stay healthy and happy,
Kathy

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

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LOOKING AHEAD

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

Winter (current) 2013: Osteoporosis, Osteopenia And Other Skeletal Issues.

Spring (May) 2013: Gastrointestinal Issues. (Submissions due March 15, 2013.) Does your G-I system give you problems? Do you use enzymes? Is one better than another for you? Have you had recurring problems with your G-I system? Tell us your experiences.

Summer (August) 2013: Motivation – What or Who Keeps Me Going. (Submissions due June 15, 2013.) Is there someone or something that keeps you motivated to live life to the fullest? Tell us who or what those motivators are and how they keep you going.

Autumn (November) 2013: Living With Pain. (Submissions due September 15, 2013.)

ASK THE ATTORNEY

Questions From the Readers, Answers From the Attorney

By Beth Sufian, JD

Happy New Year! 2013 is sure to be a special year. As I write this, we count down the days until the Affordable Care Act is fully implemented on January 1, 2013. The Affordable Care Act will make it possible for people with CF to purchase a health insurance plan regardless of their pre-existing condition. In addition, if a person cannot afford insurance coverage premiums there will be subsidies available from the government to help pay the premiums. Future 2013 columns will provide more detailed information about the Affordable Care Act and how it provides access to insurance coverage.

If you have specific questions related to insurance coverage, government benefits, education or employment rights please contact the CF Legal Information Hotline at 1-800-622-0385. The Hotline is sponsored by a grant from the CF Foundation and all contacts are free and confidential. The following are answers to questions from readers.

1. I receive Social Security benefits and want to know if I will keep the benefits forever or if the Social Security Administration (SSA) will review my case at some point.

A Social Security approval for benefits is not a "forever approval". Typically, Social Security will review the case of a person receiving either Supplemental Security Income (SSI) or Social Security Disability Insurance (SSDI) 3 to 5 years after the person is approved for benefits. However, certain things can trigger an earlier review. A person who receives SSA benefits is allowed to work part time as long as the person does not make over a certain amount of money from work activity. For a person receiving SSDI, that amount is $1,040 a month in 2013. An individual receiving SSI will have a different allowable amount to make monthly from part time work activity. That amount depends on a number of factors so it is best to contact SSA to determine how much you can make from part time work activity if you are on SSI. SSI also has certain work programs that allow a person to work and save money for a specific goal. One program is called the PASS program and may help a person with CF who wants to work and save money to pay for college or to start a business. If a person makes over the allowable monthly amount made from work activity, that person will lose those benefits. If a person is on SSDI and has Medicare, that person will lose both the monthly SSDI cash benefit and the Medicare coverage. If a person enrolls in a program called the "Ticket to Work" program and returns to full time work he loses his monthly SSDI cash benefit but is allowed to keep Medicare for 8 years. If a person is on SSI the person will lose the SSI cash benefit and the Medicaid benefits. In most states, an adult can obtain Medicaid only if he is also receiving SSI benefits. The fact that a person will have no insurance coverage without Medicare or Medicaid does not create an exception. Benefits and insurance coverage under Medicare or Medicaid will stop if a person makes over the allowable monthly work income amount.

BETH SUFIAN

Even though a person is allowed to make a certain amount from work activity, recently SSA seems to be reviewing SSA benefit recipients even if the person is working within the allowable amount. SSA is allowed to review a person's benefit eligibility status at any point in time. Some members of Congress have instructed SSA to increase their efforts to make sure that those receiving SSI and SSDI benefits are eligible for benefits. Therefore, if a person is working part time he should make sure that he would be able to provide evidence he still meets the SSA medical requirements in the event his case is reviewed. This means making sure you attend regular CF Center clinic visits and making sure you have a sputum culture performed at least once every 6 months. In addition, when discussing your health problems on the phone with your CF Center nurse, you must make sure that these issues are recorded in your CF Center clinic chart.

Lastly, in this time of Facebook, Twitter, Blogs and the Internet, be careful about what you put online

about yourself. Social Security representatives have access to the Internet and have realized that "googling" a person's name can sometimes lead to evidence that could support a termination of benefits. Use good judgment when posting or writing about yourself on line.

2. I receive SSI and Medicaid benefits. If I married my boyfriend I would lose those benefits because his income would put our household over the SSI and Medicaid monthly allowable amount. My social worker says if I just have a ceremony but do not get legally married, I will get to keep my benefits. Is this correct?

NO!! The social worker is wrong. If a person holds him or herself out as married then SSA should consider the person married and the spouse's income will be counted for purposes of determining income eligibility for SSI and Medicaid benefits. Having a "love ceremony" or a religious ceremony or any type of ceremony but not getting legally married to subvert the SSA rules is considered fraud. This activity does not result in only termination of benefits and a requirement the person pay back all benefits paid after the ceremony, but the person could also be investigated for a criminal offense. Many times a person is able to become an insured spouse under their new spouse's employer-sponsored health insurance plan, once the couple is married. So there is no reason to try to keep Medicaid and SSI and risk criminal penalties. ▲

Beth is 47 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.

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SPIRIT MEDICINE

Honesty Heals

By Isabel Stenzel Byrnes

In this Spirit Medicine article, I'd like to explore the issue of honesty. Every human being faces the moral and social implications of being honest. As people with cystic fibrosis (CF), I believe we are constantly challenged to be honest with ourselves, to be honest to others, and to be honest to the Universe or God.

Most religious doctrines start with a common dictation: *Thou shall not lie*. Lying is a cardinal sin; lying gets people into trouble, it hurts others, it breaks up families and relationships. Lying can cause much damage to ourselves. Yet, to be an Honest Abe is a bar that's often too high for most. Portrayed in the film *Lincoln*, even Honest Abe sent a white lie in a telegraph, for the greater purpose of passing Amendment 13. So why is it so hard to always tell the truth? I know I have a habit of coming up with many excuses when I am dishonest.

There are aspects of my CF experience that have made it particularly difficult to always be honest. For example, little white lies are usually socially convenient. Whenever someone asks me, "How are you doing?" I usually resort to saying "I'm fine," even if I had just coughed up blood that morning, or when I am sitting on a gurney in the ER. Was I *really* fine? These kinds of white lies are told because it's too much trouble to go into detail; or it's not their business; or you simply don't feel like opening up that can of worms. But where do you draw the line?

Every few months, I find myself sitting on an exam table, being interrogated by my nurse coordinator. She asks the usual questions: "Are you taking Fosamax? Are you taking Mycelex? Are you doing TOBI twice a day?" I find myself in a predicament. I should say yes, but why should I not tell the truth? I lose track of my Fosamax schedule. Daily pills – no problem. But once a week? Geez. That takes concentration. But, I'm nursing a broken bone now and know I am partially responsible for it. How embarrassing. I'm 41 years old and still forget this pill. And Mycelex is useless for me except for rotting my teeth. Now that I'm post-transplant, TOBI twice-a-day doesn't do anything more for me than once-a-day. Telling the truth to our health care providers can sometimes backfire; we can be labeled as non-compliant, flakey, difficult, or worse, combative. And yet, if I truly want a collaborative relationship with my care team, I feel compelled to share what works for me and what doesn't—honestly. It's not easy. And often it's the personality and warmth of the provider that makes all the difference. By being completely open, I want this provider to trust that I know my body well and I'm in control.

ISABEL STENZEL BYRNES

I've heard about families where parents do not tell their kids (older kids and teens) that they have cystic fibrosis. Clearly, every parent has a right to raise their children in the way that works best for them. But why would parents not reveal a CF diagnosis? Perhaps these parents want to spare their children the emotional suffering caused by the knowledge that they have a terminal illness. That's completely understandable. No parent wants to devastate their kids, nor introduce them into a world of anxiety, angst and fear.

In my experience, though, even as a child I was quite capable of figuring out that something wasn't quite right; and the unknown was scarier than the known. And besides, now there's the internet! To me, this secrecy or concealment is depriving these young people an opportunity to find a way, themselves, to make sense of their illness and to weave CF into their life activities and goals. Though done in the best intentions, to me, hiding CF from the patient is a disservice. It sends a message that it is not okay to talk about illness - a part of the patient's life. By thinking they are sparing their kids of suffering, these parents seem to distrust our capacity as CF patients to experience all range of emotions, to seek out what helps us feel better, and to learn to manage those emotions in a healthy way.

On a more common note for us adults with CF, I recently met a

woman who shared that none of her friends knows about her CF because she didn't get around to telling them. When she was hospitalized, she said she was "on vacation". I remember saying something like that when I was younger. While this isn't necessarily called "lying", it seems this word choice denies the chance to be authentic with who she is and what her life is about. Though every person can decide how to reveal his or her CF, I wonder what she might be afraid of. Maybe she is afraid of being treated differently, of being ostracized, pitied, or patronized because of CF.

It can be very, very overwhelming to open up to others about what is really going on. CF forces a level of vulnerability that can be terrifying. And justifiably, in certain cultures, having an illness really is a stigma, so hiding illness is a survival instinct. And yet, the energy to hide our truth is sometimes more exhausting than just letting it all out. I learned that when I was younger - big time. I was always hiding my pills, making excuses about why I needed to go to the nurse's office to do a treatment, just BS'ing at every corner with paranoia. Then I realized my own thoughts of what others were thinking were scarier than what others were actually thinking! Thankfully, I now believe being honest with my friends entrusts them with the capacity to learn to cope with having a very sick friend. We are giving those around us the opportunity to gain perspective, and more importantly, to be honest with us about *their* private lives as well.

Along the road of this CF journey, the need for honesty can sometimes become urgent. Honesty starts with admitting that you have a serious illness, or that your CF has become severe. Not everyone can do that. Denial was one of my greatest strengths. Months before my transplant, I didn't think I was so sick; I didn't think my life was so impaired by CF… but it was really a cover up. I wish I had the insight to be truly honest with myself, and see how hard it was. Therefore, being fully honest with oneself takes careful self-evaluation. I needed to stop, close my eyes, be by myself, and really, truly ask myself, "How am I?" If I earnestly listened to myself, I would NOT have said fine. I wasn't fine. That's why I crashed and had an 11th hour transplant off the ventilator. The first step to my honesty is to admit that I had real limitations. Confessing that I couldn't do certain things, like continuing to work, took courage and strength -- which I was lacking at the time.

It is easy to equate honesty with 'good' and dishonesty as 'bad'. Life isn't so simple! It is an ongoing act of maturation to face my truth and learn to articulate it to others. My focus has shifted to reasons for occasional dishonesty - shame, fear, guilt, intimidation, etc. - and an exploration of those emotions behind the truth.

As people with CF, we are constantly challenged to be completely honest with ourselves about our CF experience. To me. being honest means being real. I am who I am, and I can't change my having CF. So why deny, hide or lie about what my life is? When I'm most truthful to myself, I am more true to those around me. And telling the truth, to me, is a spiritual necessity. I lie naked before God with all that I am and have no fear; I will be understood. If I feel angry at God for letting one of my friends die, I will say it. Life with illness is hard and I will say it. When I tell the truth, it usually invites those who are compassionate to inquire more. The best people are drawn to me, and I am blessed further. ▲

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

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!

SPEEDING PAST 50...

Oh, My Aching Back!

By Kathy Russell

When I was young many people used the phrase, "Oh, my aching back!" as a way to express disgust. I remember hearing it used especially when someone would whine on about something. After a while of listening to the whining, someone would look askance at the complainer and say, "Oh, my aching back!"

So why am I saying it? I say it because my back aches. That seems reasonable to me. That begs the question, why does my back ache? Perhaps it is because I am old and have scoliosis and compressed discs. I have had spinal surgery to repair some damage in my low back, but that doesn't take away all the pain. It took care of the pain in that area, but the rest of my back still hurts.

You may be wondering why I don't "take something" to ease the pain. That's a fair question. I don't take any medicines that might inhibit my ability to breathe. Also, I cannot take any form of aspirin because I have had bleeding ulcers more than once. I'm allergic to codeine. Acetaminophen has never helped me and if I take ibuprofen for more than a day or two I start to get bruises all over. So I just learn to live with it. Besides, I am taking calcium, magnesium, zinc and glucosamine with chondroitin to try to help.

My hands and feet give me discomfort, too. It seems to be arthritis. Even when I am barefoot, it feels as if there are marbles or wadded-up fabric under the ball of my right foot, especially. The left isn't quite as noticeable. I have pain in my thumbs and index fingers. It is very difficult for me to write. My handwriting never was that great, but now it really looks like chicken tracks. I do use Voltaren® gel on my thumbs and that seems to help. Sometimes I even wear gloves to help keep my hands from hurting.

According to my bone scans, I have osteopenia in my low back but, otherwise, my bone density seems to be okay. So far, I have no osteoporosis. Spinal x-rays show that I have bone spurs in my neck and low back and compression fractures to go along with the twisting of the spine. I find that simple things such as turning my head can be very difficult on some days. I certainly cannot do "The Twist", but I manage pretty well.

I spend a lot of time in a chair that is just my size. I place a heating pad, on warm, behind my back and put my legs up on a leg rest. I put a lightweight down comforter around my legs and am very comfortable. I am able to do a lot of computer work in that configuration.

There are times when a knee or an ankle or elbow will start to hurt. I try to figure out why it is hurting, then do what I can to ease the pain. I don't expect to be pain-free, but I do like to be relatively comfortable. As a rule, my bone pains don't keep me from enjoying life. As long as I keep everything warm enough, with sox and sweaters and such, I am fine.

> *"I don't expect to be pain-free, but I do like to be relatively comfortable. As a rule, my bone pains don't keep me from enjoying life."*

KATHY RUSSELL

When the weather gets colder, as it is now, I tend to spend more time in my chair. I read, work on the computer or do jigsaw or crossword puzzles. I used to do needlework, but as I age my eyesight is not as good as it used to be and my hands don't work as well as they did, so now I don't do that.

I have no idea how much of my aches and pains are CF-related. One school of thought is that – since we have CF, everything is CF-related. Another school of thought is that – not all of our aches and pains relate to CF. I think it is a composite of both schools of thought. Some of my "arthritis" is, undoubtedly, CF-related. Some of my discomfort is from the same degenerative arthritis that both of my parents had. My sister has it, too, and she doesn't have CF.

That raises the specter of carrier symptoms. Could my sister's pain be because she is a carrier? Since she has never been tested for CF, we don't really know. Could it be that we all just have arthritis and it has nothing to do with CF? All I know is that it can be a real pain in one's bones.

My pain started in my knees and ankles, when I was about 12. My 4-H group had roller skating parties on Thursday evenings. On Fridays, my legs did not want to work. My legs would ache so badly and my knees would get red and tender. I would stay in bed for a day or two and all would be okay again. I took dance classes two days a week and didn't get negative effects from that. Perhaps the added weight of the skates had something to do with the pain. After a while, I stopped going to the skating rink. It just wasn't worth the pain.

As I sit here typing this, I am comfortably warm. I can look out and see the trees and the sky. I feel that I am still a part of what is going on, even though I am not up and running around. I am satisfied at this stage of my life.

If I were younger, would I feel satisfied? That is a tough question. I used to work through some of my pain. I was young enough that I could fool myself. If I stayed as busy as I could, I was too tired to notice the pain. On rare occasions, the pain would get the best of me. Then I'd have to stop, assess my situation and make some adjustments. I didn't like it, but I was able to keep going that way. Perhaps that wasn't a wise way to handle it, but it allowed me to work and to play. I tried to stay very active in hopes that the activity would keep my joints working better and longer. I feel that it worked. It is only the past ten years or so that have found me less able to do all that I want to.

At times I have had physical therapy with exercises and massage to ease my back pains. The massage did relax my muscles and that eases some of the pulling on my twisted spine. The exercises will help for a period of time, then they seem to help less. I think that this is my "new normal".

So that means no tap dancing for me. That's okay. I couldn't tap dance when I was younger. No more running up stairs, but I had to give that up when I got supplemental oxygen. My portable oxygen concentrator (POC) weighs enough that I can't carry it for more than a few steps on level ground. At home, where I have a larger concentrator and about 50 feet of hose, I have no stairs so I don't have to deal with that.

I am very careful about keeping track of my oxygen hose. It has tripped me in the past and I have no doubt that it will try to do it again. Both Paul (my husband) and I are very aware of that slithery hose and try to avoid it. When I was tripped by the hose, a couple of years ago, I hurt one knee. It took over a year for my leg to feel okay. I do not want to go through that again.

One way that I get some relief from back pain is that Paul rubs my back every night before bedtime. He scratches it delicately then rubs it a little more vigorously. The scratching eases a lot of the tightness and the rubbing relaxes the muscles. I find that I am able to sleep much more comfortably after his ministrations.

We have taken some steps to make sleep a little more restful. We purchased a mattress that is memory foam. We have had it more than ten years and find that it still is the most comfortable sleep either of us has ever had. We use a down comforter in a duvet for warmth. It is light enough that it causes no discomfort from weight.

I got an all-natural pillow. It is made of cotton with a wool fleece and millet filling. There are no dyes or perfumes used. The millet is sterilized and in its own compartment. I can adjust the amount of millet to meet my comfort needs. The fleece is in a special section on the top of the pillow cover. This pillow doesn't hurt my ear where the oxygen hose is and it conforms to my head for maximum support. I have less shoulder and neck pain since I have been using this pillow.

I mentioned that I have trouble with my feet. To help keep them from hurting, I use prescription orthotics. I buy shoes that can hold the orthotics and wear this added support all the time. My feet, ankles and knees have felt much better for the years that I have been wearing the orthotics. Obviously, I don't wear high heels or high-fashion shoes, but I haven't worn such things in decades, so I feel no loss. I figure that anything that eases the discomfort in my feet is a good thing, and I always have liked European style clogs anyway.

So, to sum up, I have a good mattress, good pillow, comfortable and supportive shoes, a comfortable chair that fits me, clothes and covers to keep me warm, and the good sense to be careful when walking around my oxygen hose. I hope that each of you who have bone and/or joint pain can find what works for you to keep your life livable. I don't want you to have to say, "Oh, my aching back."

Stay healthy and happy and may 2013 bring you good health, love and laughter. ▲

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

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

Andrea Eisenman
New York, NY
48 on November 28, 2012

Delayne Santos
Gulfport, FL
45 on February 14, 2012

Carol Shepherd
Ft. Worth, TX
55 on October 18, 2012

Erin Phillips Taylor
Groveland, FL
28 on August 2, 2012

Laura Tillman
Northville, MI
65 on December 22, 2012

Wedding

Christine & Matthew Conway
Arlington Heights, IL
4 years on November 29, 2012

Delayne & Gerry Santos
Gulfport, FL
20 years on September 19, 2012

Carol & Robert Shepherd
Ft. Worth, TX
31 years on July 11, 2012

NEW BEGINNINGS

Wedding

Jamie Crane & Kristen Koch
Baton Rouge, LA
Married on October 6, 2012

Birth

Christine Conway, 29
Arlington Heights, IL
A daughter, Penelope Rose
On September 3, 2012

Transplants

Jamie Crane, 26
Baton Rouge, LA
Bilateral lungs
March 29, 2012

Erin Phillips Taylor, 27
Groveland, FL
Bilateral lungs
July 31, 2012

Information from the Internet...

Compiled by Laura Tillman

PRESS RELEASES

Aradigm announce encouraging data from cystic fibrosis study

Aradigm Corporation has announced findings about the anti-inflammatory effects of the company's inhaled ciprofloxacin. Aradigm's drug, Pulmaquin, is a mixture of a small amount of free ciprofloxacin that provides a spike of immediately available antibiotic for rapid anti-infective activity plus liposomal ciprofloxacin that provides sustained release of the antibiotic in the lung, facilitating once daily dosing and improved airway tolerability.

The company's once-a-day novel inhaled formulations of ciprofloxacin are encapsulated in liposomes, allowing for a sustained release of the drug within the lung and improving airway tolerability. The formulations are to be used for chronic maintenance therapy as they are expected to achieve higher antibiotic concentration at the site of infection and relatively low systemic antibiotic concentrations to minimize side-effects.

Lipoquin is a liposomal formulation of ciprofloxacin. Pulmaquin is a dual release formulation that is a mix-

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Mailbox

I have been subscribing to *CF Roundtable* for quite a few years now and I thought people might be as encouraged by my daughter's milestones as I am by reading other people's milestones. Thank you and I want to take this opportunity to tell you how much I enjoy this publication.

Rosa Judycki
Schaumburg, IL

Hi! My name is Brandi Edelman and I am an adult living with cystic fibrosis and diabetes. I received a copy of *CF Roundtable* from my CF center, at my last hospitalization, and thought what a great source of information it was! Then I thought...hey, why don't I write an article? I feel living with CF is so serious a lot of the time that I wanted to add some humor to it, just to make someone smile! I am, by far, not an author/writer, but I thought I would give it a shot.

I am a very blessed 32-year-old, who recently got married for the first time to a terrific Kansas farm boy, have three wonderful stepchildren, and work as a chiropractic assistant for a great doctor. I am surrounded by four-legged family members that are my salvation when times get tough (there is nothing like the love and loyalty of a dog.) I also have a tremendous support system in my parents and my in-laws for which I am thankful.

Brandi Edelman
Sabetha, KS

I'm a 57-year-old with CF in Texas. I was misdiagnosed until I was 28 years old. I worked in corporate communications for major corporations in Dallas for 25 years. I've been on disability since 2007. I am in the process of learning to paint watercolors, which I really enjoy.

Joan Hammond
Marshall, TX

Heartfelt thanks for the ever ongoing wealth of information and the inspirational messages of love and hope.

Sincerely,
Phyllis Kossoff
New York, NY

As a long time caregiver for patients with CF I very much enjoy reading *CF Roundtable*. I trained at Children's Hospital of Philadelphia in Pediatric G-I with Guilio Barbero and then returned to Akron in 1967. At that time we had 20-plus patients with the mean survival of 8.5 years. Now we have 230-plus patients with a mean survival of 38.5 years. What an exciting trip!! I retired three years ago but continue to work at the Hospital and am a strong advocate for CF in the community. Keep up the good work.

Robert T. Stone M.D.
Akron, OH

On October 18, 2012, I turned 55 years old! This is a milestone to me! I had my 31st wedding anniversary on July 11, 2012. We are surviving with CF!

Carol Shepherd
Ft. Worth, TX

You have a wonderful newsletter.

Thank you,
Nora Furey,
Mom of 37-year-old son [who is] married
with a four-year-old and newborn twin boys!!!

Thank you for your investment in CF education and communication for the cystic fibrosis community. *CF Roundtable* is such an important and valuable resource for all those affected by CF.

Anonymous

Thank you so much for your wonderful newsletter! I really appreciate all the hard work you all do. I especially read with interest the stories about transplant because I have been accepted at Barnes in St. Louis. I'll be officially listed after January 1, 2013. I am so ready! Keep up the good work!

Johanna Libbert
Richland, IN

Thank you for your informative and inspiring articles.

Marie McCloskey
Wynnewood, PA

Thank you for the Focus [topic] in the last *CF Roundtable* regarding lung transplants. It came at a good time for me as I had just been referred to a transplant specialist in October. I'm sure the confusion mixed with hope, depression and anxiety that I felt is normal. The inspiring articles gave me some real answers and optimism. I see the transplant specialist in two weeks again to decide the timing of testing relative to my overall health, age and lung functions. I look forward to *CF Roundtable* every time.

I wish you and the contributors a very happy and healthy New Year.

PS. I was excited to learn that Houston will be hosting the Transplant games in January 2014. Who knows, maybe I'll be a participant.

Bonnie Bleiweiss
Houston, TX

Voices from the Roundtable

Do You Want Fries With That? Standing up for yourself…

By Brandi Edelman

Non-compliant. Ornery. Opinionated. These are just some of the words that I, as a CF patient, have been called (along with a few others I am sure would not be allowed in this respectable publication!). And I, as that said CF patient say, "Thank you," because I AM all of those things.

I was diagnosed with cystic fibrosis (CF) at 5-months-of-age and have never known life without treatments, medications, and hospitalizations. And, like many of you, I know my body. I know what medications work for what ailing issue I may have at the time: I know what I can realistically do on a day-to-day basis regarding workouts and treatments. I know that my current CF team has my best interest at heart. With that said, despite the many doctors I have been to throughout my years (and in many locations all over the country), I find that the best advocate for my healthcare is ME.

At 13 years of age, I decided after leaving two CF visits in tears, that I would never step foot back in the building where it was located – ever again. With full support from my parents, I started seeing a pulmonologist who actually listened and cared about what I had to say. He ultimately diagnosed me with diabetes. You talk about someone in a bad mood - try living with blood sugars over 400 and going through puberty! I knew something wasn't right with my body and it took me standing up for myself and getting the help I needed, outside of a CF center, to make things better.

Fast forward a few years and my family moved to the Midwest, where I once again started over with new doctors. I decided to go back to a CF center, as it came highly recommended. Times were good. I liked my CF team and my English accented doctor was awesome. You have to love a doctor who comes in on his Sunday off, in full motorcycle gear, to check and see how you are doing!

> "I know my body. I know what medications work for what ailing issue I may have at the time."

At this time, the CF center was introducing an "adult" program and looking for young prey, I mean patients, to move up and get things started. Although hesitant to leave my wonderful surroundings at the pediatric clinic, I agreed to help out. The first visit with my new "adult" CF doctor left me yearning for happier times with the Englishman!

The doctor came in with a bit of a swagger, one I recognized well and I thought to myself…here we go. We went through my history, the normal pleasantries, and what I had going on with my health at that particular time. I told him what I like for medication to fix the issue and then he said something that made my mouth drop to the floor. He looked me straight in the eyes and asked me if I wanted "fries with that" (meaning my "order" of medication).

What did he just say to me? Too stunned to answer, I simply sat there. When the nurses came in to finish up my appointment, I let them know that he was not to talk to me like that and I did not appreciate it! Leaving, fuming under the collar, I thought that he won't last long working with CF patients.

Our time on earth is short and precious and we don't have time and patience to deal with rude doctors! I made my follow-up appointment after I had had time to settle down and thought I would give Dr. Swagger another shot. He came in humbled and apologetic at my next visit, which I appreciated, and I decided to forgive. I laughed, actually, because I had guessed that his short three months of working at a CF center had showed him that we who have CF may not have M.D. after our names, but we know things and it is not our first rodeo!

Dr. Swagger and I are going on 12 years in our relationship, or what I like to call our "health marriage". We have had several issues that have come up, but he respects my opinion and I respect his knowledge and advice. We look back and laugh at the "do you want fries with that?" comment. He tells people how "mad" I was and I tell people how "arrogant" he was and we all have a good laugh.

I am now 32 years old and doing well. The scary word "transplant" was brought up at my last hospitalization. Dr. Swagger and I had a nice visit about it, and I told him that God will let me know when I am ready. But, at this point, I am living life to the fullest and I will be the one to make the decision when the time comes. And when I do, I am sure it will be a bit ornery, maybe a little opinionated, with a hint of non-compliance. ▲

Brandi is 32 and has CF. She lives with her husband and children in Sabetha, KS. You may contact her at: brandirae90@yahoo.com.

age. Short children and children with delayed puberty will have bones that appear less dense than normal even though they have normal density.

According to the World Health Organization, osteoporosis is defined as bone mineral density standard deviation score (BMD-T) <-2.5; a bone density in this low range predicts future fracture risk. In children and adolescents, the use of a BMD-T is inappropriate and a BMD-Z must be used. How well BMD-Z predicts fracture risk in younger individuals is less well established. Thus, osteoporosis is defined in children and younger adults as BMD-Z<-2 and a history of significant fractures, including long bone fractures of the upper and lower extremities and compression fractures of the spine.

Young adults with cystic fibrosis are at increased risk of spine and rib fractures. These types of fractures are particularly problematic in cystic fibrosis because the pain and deformity they cause can interfere with breathing and appetite. As one might expect, these same individuals have decreased bone mineral density. Bone density appears relatively well-preserved in childhood but accrual during puberty is less than expected. In adults with CF, there is bone loss. Essentially, in at least some individuals with CF, optimal bone density is not achieved during puberty and the bone loss of aging occurs earlier. Much of this data is derived from individuals who were children, teenagers, and adults at least ten years ago. The outlook for younger individuals may be more favorable. Moreover, little data is available in older adults with cystic fibrosis in whom aging may compound the risks for decreased bone mineral density and increased fracture risk or in individuals who have had transplants in whom anti-rejection medications may compromise bone health, particularly in the first year after transplant.

The reasons for poor bone health in CF are numerous:

- Glucocorticoids (for example, prednisone) decrease bone formation
- Decreased sex steroids increase bone loss
- Poor nutrition and malabsorption can lead to deficiencies in vitamins important for bone (D, K, Zinc, Ca)
- Poor nutrition, lack of exercise, and decreased weight-bearing activity can lead to decreased muscle—the pull of muscle on bone is important for bone strength
- Inflammation can also increase bone loss
- Renal disease can increase bone loss

Suggestions for DXA

The 2005 CF Foundation recommends obtaining DXA in

- Adults >18y
- Children >8y at increased risk for decreased bone mineral density
  - Ideal body weight <90%
  - FEV_1 <50% predicted
  - Prolonged glucocorticoid use
  - Delayed puberty
  - Previous fracture history

Individuals with CF will require serial DXA evaluation depending on their BMD-Z or BMD-T. If the DXA is normal (BMD-Z/T>-1) then the DXA can be repeated in five years (sooner if concerns arise). If the DXA BMD-Z/T is between -1 and -2 the DXA can be repeated every 2-4 years (again, sooner if concerns arise). A yearly DXA is recommended for DXA-Z/T <-2. DXA should also be obtained pre-lung transplant. The frequency post-transplant is less clear. Given the risk of decrease in BMD in the year after transplant—a year following transplant is likely prudent with the subsequent intervals dictated by previous results, interventions for bone, and overall clinical status.

The CF Foundation also recommends annual screening for vitamin D deficiency. Renal disease, not uncommon with repeated intravenous antibiotic use and in the setting of transplant, can impact vitamin D metabo-

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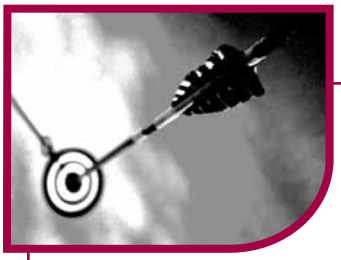
CLUB CF ONLINE

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.



FOCUS TOPIC

OSTEOPOROSIS, OSTEOPENIA AND OTHER SKELETAL ISSUES

Bone Business

By Anne Williman

I guess I shouldn't have been surprised when I developed osteoporosis. After all, both my parents have it, and I knew there was a tie-in with my having cystic fibrosis.

Probably ten or fifteen years ago, my CF doctor asked me to get a DEXA scan, which measures the bone density at the hip and spine. When I went to get the test, I was pleased to find out it was non-invasive. (I appreciate anything that doesn't involve pain!) You just lie there and are moved into the machine for a few seconds.

When the results came back, I did have low bone density. So the doc started me on Actonel. This oral drug is taken once a week when you first get up in the morning, before eating or drinking anything. You take it with a big glass of water and then have to wait half an hour in a sitting or standing position without eating.

I was not fond of taking it, though I spent my half-hour reading. Still, it seemed like Thursday, the day I took it, came too often. It did seem to help my bone density though.

A few years later, I heard about Boniva, and I asked the doc if I could switch. He agreed. This drug was better since you only had to take it once a month. You did have to wait an hour afterwards, but I figured I could do that when it was that infrequent.

I wasn't on it too long before I started having trouble with acid reflux. To control it, I ended up trying several different drugs and not having much success until I got on Nexium twice a day.

That meant the end of my days on oral osteoporosis drugs, because you can't take the meds with acid reflux. I asked the doc about other drugs.

Turns out he had a patient who had had good success with Fortical, a nasal spray. One spray a day, alternating nostrils, was all it took. So I went on that.

My next DEXA scan didn't have the huge improvement that his other patient had had, but the doc was satisfied that it was working. I was satisfied because I liked taking it far more than the oral drugs.

> "I try to be more careful about not putting myself in positions that might result in a fall."

I was on Fortical for years, and then in September 2010, I got seriously ill with pneumonia. I ended up spending two months in the hospital, 30 days in ICU, and 10 days on a ventilator. It took a long time to recover from that; in fact, two years later, I am still not back to where I was physically before that fall.

But when I had a DEXA scan the summer afterwards, my results were not good. The bones were worse. The doc thought I should go on Reclast, a drug taken by IV. That was not appealing; I get enough IVs as it is.

"Couldn't the bone loss be the result of my being so sick?" I asked.

He agreed that could be the problem and was willing to let me stay on Fortical another six months before retaking the test.

When that time passed and I had my next DEXA scan, I was pleased to see there was improvement. The doc was fine with me staying on the nasal spray.

I'm still on it today, and so far my bones seem to be doing pretty well.

I do try to do other things to prevent bone problems, like work hard to avoid falls. So far I've only had two, both of which didn't result in any serious injury. The first was shortly after I got home from the hospital after the pneumonia. My husband helped me out onto the front porch to enjoy a few minutes of the warm fall weather (I had basically missed all fall in the hospital.) But when we started back in, he went ahead of me, not realizing that I did not have the strength to lift my foot up the step. (That's what two months in a hospital bed does to you.)

I ended up falling backward, cracking my head on the cement porch. It hurt for a few minutes but did nothing more.

Just six months ago, I had another fall, in which, for some reason, both of my knees decided to give out at the same time. (I guess old age is setting in!) I ended up on the floor, but again was not really hurt.

After those experiences, I try to be more careful about not putting myself in positions that might result in a fall. I also make an effort to walk our dog outside as often as possible, since I know exercise is good for my bones, not to mention my lungs.

I have always consumed a lot of milk and dairy products, and I continue to do that for the calcium. I prefer skim milk, even though I know

I need the calories in whole milk. That's because when I was growing up in the '60s, people with CF were put on a low fat diet. I was not allowed to eat fatty foods like bacon, peanut butter, or whole milk.

Now, of course, it's been proven that people with CF need those extra calories. But I just can't get used to the taste of whole milk, so I continue to drink skim. (I do enjoy bacon and other fatty foods though.)

So for now, the osteoporosis isn't a major concern for me. (Wish I could say the same about the CF and the CFRD.) I take the DEXA scan (remembering I can't wear my jeans that day since the metal from them would show up in the picture), and use the Fortical, and pray for the best.

I pray I won't have a spontaneous spine break like my mother did from her osteoporosis, sending her to the hospital and nursing home for months. I pray I won't have another fall but, if I do, I pray I won't break a bone. Beyond that, there's not much I can do. I'm thankful for the good days, the parts that work the way they should, the small improvements that come. And I deal with the things that don't go as planned. Isn't that the way it usually is with CF? ▲

Anne is 59 and has CF. She lives with her husband, Jon, in Middletown, OH.

TILLMAN *continued from page 10*

ture of Lipoquin with unencapsulated ciprofloxacin.
http://tinyurl.com/9nxers8

Aradigm Corporation: Virginia Commonwealth University Scientists Report Anti-Inflammatory Effect of Aradigm's Inhaled Liposomal Ciprofloxacin

Ciprofloxacin is a widely prescribed antibiotic to treat infections of the lung frequently experienced by cystic fibrosis (CF). It is often preferred because of its broad-spectrum antibacterial action. The company's once-a-day novel inhaled formulations of ciprofloxacin are encapsulated in liposomes, allowing for a sustained release of the drug within the lung and improving airway tolerability. The formulations are to be used for chronic maintenance therapy as they are expected to achieve higher antibiotic concentration at the site of infection and relatively low systemic antibiotic concentrations to minimize side-effects. Lipoquin is a liposomal formulation of ciprofloxacin. Pulmaquin is a dual release formulation that is a mixture of Lipoquin with unencapsulated ciprofloxacin.
http://tinyurl.com/9kebemj

Cystic fibrosis disrupts pancreas two ways in CF-related diabetes

A University of Iowa study suggests there are two root causes of a type of diabetes associated with cystic fibrosis (CF). Using a ferret model of CF, the study shows that CF progressively damages the pancreas, disrupting insulin production. The study also found that CF disrupts the pancreas' insulin-producing islet cells from birth, well before the physical damage occurs.
http://tinyurl.com/8zgqmwj

Vertex Reports Third Quarter 2012 Financial Results and Recent Progress in Development Programs

- Ongoing global launch of KALYDECO
- Efforts to help more CF patients with ivacaftor monotherapy
- Combination therapy for people with two copies of the F508del mutation
- Research to identify additional CF treatment regimens

http://tinyurl.com/cy9nt2h

Potential Treatment For Cystic Fibrosis Lung Disease

Vancouver researchers have discovered the cellular pathway that causes lung-damaging inflammation in cystic fibrosis (CF), and that reducing the pathway's activity also decreases inflammation. The finding offers a potential new drug target for treating CF lung disease.
http://tinyurl.com/aa4oq4v

Fighting bacteria with mucus

Results from a recent MIT study suggest a possible new source of protection against biofilm formation: polymers found in mucus. The MIT biological engineers found that these polymers, known as mucins, can trap bacteria and prevent them from clumping together on a surface, rendering them harmless.
http://tinyurl.com/d4buk2d

Ecological approach could help cystic fibrosis sufferers

Bringing the methods of microbial ecology to bear on lung infections in people with cystic fibrosis may yield insights that will radically improve their lives and help them survive longer. The idea is that we need to look at the bacteria infesting patients' lungs not as a single homogenous invading force but as a whole ecosystem of different species, interacting with each

Continued on page 16

Ask My Bones How They Feel

By Nicole Matthews

At the young age of twenty-four, I feel as if I were eighty years old on some days. A few years back I was diagnosed with rheumatoid arthritis (RA). I was diagnosed with scoliosis around middle school age. I remember when the girl who took my bone density scan four years ago asked me how old I was, because I am really not in the "correct" age for getting this test done. For a CF patient, another test is simply life.

I knew I needed to figure out my pain in my joints. When the weather changes here in Buffalo, NY I can surely feel it in my joints and deep in my bones, especially when it's super damp and cold. But to help with the inflammation, I simply just take a nice warm shower, take some Motrin, and try my hardest to be active.

I know some days it's really tough to be active due to not being able to move and the pain being horrible. But I know that moving helps healing… sometimes. Other times I think I push myself a little too hard.

The weird thing is when I was able to eat gluten (found in wheat, rye, barley, modified food starch and oats) my bones hurt more often. Since I went gluten-free, three years ago, my bones do not inflame as much. I truly believe cutting gluten out of my diet has helped my RA, and the pain in my joints.

NICOLE MATTHEWS WEARING A MASK.

I still have bad swelling in the morning in my feet and hands, but the pain is nowhere near what it used to be. I also use heating pads when needed, mainly on my back. I don't know about other CF patients, but I constantly have an achy back, maybe from coughing, or from the "barrel chest"; but sitting up straight and stretching out a lot seems to help. I also get pain in my chest when I am coming down with something, usually from coughing so hard, but keeping up on treatments and medications helps.

The big thing with all these different problems is to try and stay positive, and try to think of a better tomorrow. Life is like a rose – there are thorns – but it still is beautiful. ▲

Nicole is 24 and has CF. She lives in Orchard Park, NY, with her boyfriend, Michael, two cats and a hedgehog. She may be contacted at: abnormalnicole@gmail.com or on Facebook.

TILLMAN *continued from page 15*

other and inhabiting diverse niches. We have found that a CF lung is a more diverse and complex ecosystem than previously realized, containing hundreds of unique bacterial species. As a patient's lung function declines, the diversity of the microbial community in their lungs also drops. It's possible that the latter is causing the former. Ultimately this could point to ways to help patients by being able to predict worsening of disease by using the bacterial communities as marker for the state of the disease. Researchers already knew that the lungs of CF sufferers are often dominated by a few key species of bacteria, and have succeeded in growing these in petri dishes from samples of sputum. But this approach has serious limitations. There are many kinds of bacteria living in the average infected lung, and each needs different conditions to flourish. Growing one kind may need warm, moist conditions with plenty of oxygen; another may depend on an oxygen-free environment. Unless you know what's there already, it's impossible to grow every species present because you don't know what growth conditions to provide. So growing bacteria isn't a viable way of finding out what's there. Instead, the team has been using high-throughput genetic sequencing that lets them take a com-

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PHOTO BY STEPHEN BOYER

Vrksasana

Magnificent tree
reaching upward toward the sky
grounding me

Miraculous breath
flowing throughout my body
sustaining me

Mind, body, spirit
seeking balance and wholeness
strengthening me

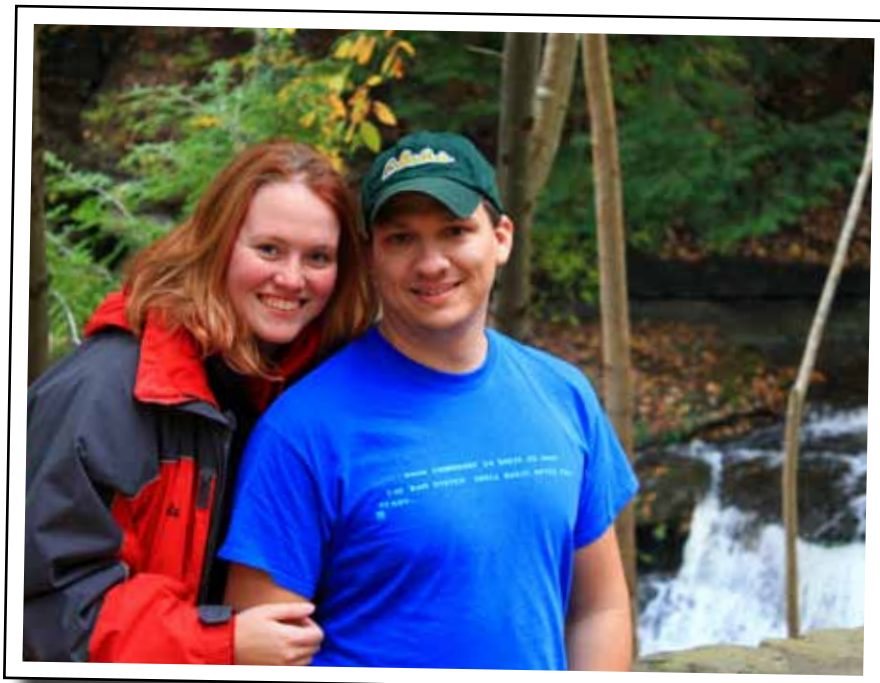
-R. Petras, 2000

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

RANDY AND STEPHANIE RATH WITH THEIR DOGS, ROCKY AND ROSIE, IN THEIR GARDEN. IN LOWER LEFT OF PHOTO, A STATUE OF A PIG WITH WINGS, STEPHANIE'S INSPIRATION FOR HER BLOG.

BRANDI EDELMAN



NICOLE MATTHEWS AND MICHAEL KOWAL AT LETCHWORTH STATE PARK, NY, COLUMBUS DAY, 2012.

CHRISTINE AND CHRIS KVAM WITH THEIR 48-POUND LAP DOG, HILDY.

CYNTHIA DUNAFON HIKED FROM THE SOUTH RIM OF THE GRAND CANYON TO THE COLORADO RIVER (AND BACK). AFTER A NEARLY 5,000 FT DESCENT FROM THE RIM, SHE IS STANDING ON THE BRIDGE OVERLOOKING THE COLORADO RIVER – HAPPY TO BE FREE OF HER BACKPACK AND AMAZED AT THE INCREDIBLE BEAUTY OF THE INNER CANYON.

Debbie Ajini
August 30, 1970 – October 26, 2012

In *CF Roundtable* (Autumn 2012), Debbie Ajini wrote an article regarding the bilateral lung transplant she received on July 28, 2012. It was full of life, hope, joy and promises of all to come. Debbie was doing well and enjoying her new found freedom from the use of supplemental oxygen and the constant exhaustion of trying to do anything that required the least bit of effort. Shortly after writing the article, and prior to publication, Debbie developed a serious infection. She had to undergo another surgery. Unfortunately, despite Debbie's spirit, spunk, and feistiness, she was not able to survive yet another assault on her body and died three days after that surgery on October 26, 2012.

Debbie touched many lives during her 42 years on earth. She had friends far and near and was a source of inspiration. She spent many years on the board of a CF organization in her local area, helping it to evolve from a camp for those with CF to an organization that provides financial help in the form of educational scholarships, camp scholarships, transplant expenses, and many other forms of support for those in need.

In 2006, Debbie was elected to the USACFA board of directors. She served as a Director of USACFA until her death. Also, she served as Secretary of USACFA for one year. She wrote a column called "A Deep Breath In" for *CF Roundtable* and she was a regular on the USACFA blog. She is survived by her husband, Louie, and innumerable family and friends. Debbie will be missed.

TILLMAN *continued from page 16*

plete census of all the bacteria in a sputum sample, as well as getting an accurate sense of how abundant each one is.
http://tinyurl.com/cwqw8e7

Savara Pharmaceuticals' AeroVanc Granted U.S. Orphan Drug Designation for the Treatment of MRSA Lung Infection in Cystic Fibrosis Patients

AeroVanc is the first inhaled antibiotic being developed to address the growing population of MRSA-infected CF patients. AeroVanc™ (vancomycin hydrochloride inhalation powder) is a proprietary inhaled dry powder form of vancomycin in a capsule-based device designed for convenient self-administration. By delivering vancomycin directly to the site of infection, AeroVanc is expected to improve clinical efficacy and reduce adverse effects due to systemic drug exposure. AeroVanc has demonstrated positive safety and tolerability results in Phase I clinical studies with a pharmacokinetic profile that supports its potential as a once- or twice-daily treatment for pulmonary MRSA infection.
http://tinyurl.com/c93ph4n

Aptalis Pharma Announces U.S. Commercial Availability of VIOKACE (Pancrelipase) Tablets

Aptalis Pharma announced that VIOKACE (pancrelipase) Tablets are now commercially available in the U.S. Approved by the U.S. Food and Drug Administration on March 1st, VIOKACE, in combination with a proton pump inhibitor, is indicated in adults for the treatment of exocrine pancreatic insufficiency due to chronic pancreatitis or pancreatectomy.
http://tinyurl.com/bvdprcc

First phase II study of combined VX-809/ivacaftor in F508del. Report From the 26th North American

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Voices from the Roundtable

Living with CF

By Chris Kvam

What does it mean to fight CF? This phrase, which I see and hear all the time, has lost its meaning for me. I can understand the desire to label CF as "enemy" and something that must be beaten. "Fighting CF" may be an excellent way to raise money for the development of new treatments without which I cannot "win" and for which I am deeply indebted. However, clinically speaking, I feel that "fighting" a long-term chronic illness such as CF is inherently unsustainable and contributes to serious problems such as anxiety and depression in the long term which may, likely, manifest in non-adherence and negative clinical outcomes.

While using language such as "fight" and "win" in instances of acute illness or the sudden onset of disease, such as cancer, may motivate or even scare individuals in the short term into treatment and adherence, is it helpful for individuals with conditions such as cystic fibrosis? I do not believe that it is. For example, I was diagnosed with CF at age 4. I have not lived a day without CF, and my very DNA is an expression of the condition. Who is the enemy? What am I trying to beat? How do I know if I "win"? Can I "win"? Cystic fibrosis is a progressive, chronic illness. There is no restoration to a pre-disease state. The best I can hope for is a maximization of my potential lung function at any one point-in-time.

CHRIS KVAM RIDING IN THE 2012 HIGHLANDER CYCLE TOUR.

I am extremely adherent to the comprehensive treatment regimen that I have developed with my CF care team. Over the last ten years, my lung function has declined about 10 percent. Am I "losing"? Perhaps. Am I not doing enough? As I sit writing with a neb cup in my mouth, flutter valve at my side and a PICC line in my arm, I think not. What about the individuals struggling to keep their PFTs in the 40s? Or facing lung transplant? Have they lost? Are they losing? Is it helpful to define their experiences in such terms, or does doing so contribute to levels of stress and fear, erode sense of self and contribute to continued decline? I am not a clinical researcher, but how can it not?

So, if I'm not fighting CF, what am I doing? I'm living with CF. Growing up with CF has given me a drive to live as fully as possible, and has helped me distill what is truly most important to me. I set attainable short- and long-term goals in many areas of my life that motivate me to adhere to my treatment regimen. If my health were to significantly decline, I know that while I could stand to lose the ability to do many of the things I love, I also know that nothing could take away what I have already done. ▲

Chris is 32 and has CF. He is married, is an avid cyclist and is an Assistant District Attorney. He lives in Rochester, NY with his wife, Christine, and a 48-pound lap dog, Hildy.

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COUGHING WITH A SMILE...

What 'SUP?

By Jennifer Hale

Hello *CF Roundtable* readers! I hope the holidays are being kind to y'all and you are enjoying the season. I recently had the opportunity to be interviewed by a fellow person with CF here in Florida about Stand Up Paddle Boarding. It is a sport I recently was introduced to since moving from snowy Chicago to sunny Florida. I thought it would be fun to share with y'all the article we wrote together. The following paragraph was expertly written by my friend, Chris. I hope you find the following Q & A fun and interesting! Enjoy!

SUP (stand up paddle boarding) is the latest international fitness craze to take center stage. In all fairness, paddle boarding has been around since humans were using long sticks to push themselves around on floating logs; and while today's paddle boards may be more advanced, the same 'primitive' concept remains at the heart of the sport. SUP has really evolved in recent years and branched off to include recreational paddling, flat water racing, open water racing, surfing, and yoga. What makes SUP so appealing is the sport's ability to accommodate people with or without disabilities- at all skill levels- in a wide range of water conditions. People can stand, lie supine, kneel, and sit (on the board or on an inflatable seat attachment) as they paddle with their hands or use a single or double blade paddle. One can even add small sail attachments to get a little help from the wind. It's the versatility of SUP that allows it to compliment a variety of therapies that are beneficial to children/adults with cystic fibrosis. With such great potential as a therapeutic tool, there's no wondering why Cystic Fibrosis Surfing/SUP Experience Days are now sponsored throughout Florida and other parts of the United States. I recently had the opportunity to catch-up with Jennifer Hale, a 40-year-old person who has CF and CF-related diabetes, and chat about how she benefits from SUP.

C: Aloha Jennifer.

JH: Hello readers out there! What'SUP ? How are you? LOL.

C: How did you get into SUP fitness?

JH: I am new to this sport and new to sunny Florida. I hail from the Windy City, so the only board I was familiar with was the one that rode on snow and down a mountain. Then I took a SUP class out on the gulf of Mexico, when I first moved to Florida, and fell in love with the sport!

C: What do you find most appealing about SUP?

JH: When I moved to Florida I just had to get out on that water! Stand Up Paddle Boarding is a great sport for all ages, genders, and disabilities. SUP is a board that looks like a surf board and you can stand, kneel or sit on it and paddle with an oar out into the water. The three aspects of it that I like the best are: ease of doing it, breathing salty air and the spiritual aspect of being out on the vast open space of the water. I really like to be out on the open water. It is really spiritual and peaceful for me.

> *"Stand Up Paddle Boarding is a great sport for all ages, genders, and disabilities."*

JENNIFER HALE

C: Is there a particular reason why you prefer ocean conditions over freshwater conditions?

JH: Being out on the water you get all that natural hypertonic saline solution. Got to love medicine that comes naturally from Mother Nature. The salt in the water and, thus, in the air is really good for our lungs! I do not mind ocean or freshwater. What I do like is calm waters. When the water is calm, like out in the mangroves, it is easier to balance vs. when I am out in the gulf; the waves make it a different workout. So, it just depends on the kinds of conditions one prefers and the kind of workout one is looking for with the SUP.

C: I'm all for taking in free hypertonic saline treatments while enjoying a sunny day at the beach! Even though

Florida waters are traditionally flat during the summer months, there is still small wave action in the Atlantic and Gulf waters. I would imagine launching your SUP board from the beach would be a challenge. How do you do it?

JH: You just have to go on the board like a surfboard, lying on your stomach, to get over the waves. Or you can just walk out into the water holding the board to your side and walking beside it. Once you get out past the waves you can lie on your stomach and paddle out to the more open water and then get your bearings to sit and/or stand up on the board. Launching from the gulf is more challenging than launching from a mangrove area because the gulf has waves vs. flat water. All in all, paddle boarding is very easy to do; and if you cannot stand on it then you can sit. So you are just floating around on this board and using your oar to paddle around. Once you get the hang of it you can stand up, which then will work your core muscles and use balancing techniques. What I like about this is if you are too tired it is easy to just sit and paddle on it like a canoe. I also like that you can hook some bungee cords to hold a little cooler for snacks and drinks. Having CFRD, it is important for me to have access to juice and with the depletion of salt from working out in the sun it is important to have those Gatorades handy. Even though I am out in the middle of the ocean I can still have my cooler with goodies attached to the board. Sitting or standing, it is a great workout and not too strenuous if you do not want it to be that way. You put into it what you want to get out. Paddle more or paddle less. Sit or stand. It is a fun workout.

C: With a vast majority of SUP boards longer than 9 feet, do you find SUPs challenging to transport on land and maneuver on water?

JH: I have not tried many lengths of boards on the water, so I'm not sure what the different lengths "feel" like out on the water. I can say carrying the boards is not as bad as it appears to be. They are lighter and have a space to put your fingers so you can carry the board. Being only 5'1", maneuvering a 9-foot or longer board is a workout in itself. The boards are not as heavy as they appear and there are shoulder straps and carts that you can purchase to transport your board. It's something to think about due to the size of the board relative to the person's size; and the condition of the person's health will factor in when deciding how to transport the board from land to sea.

C: What are your thoughts about using the paddle board to enjoy Florida's wildlife in and around our freshwater lakes, rivers, and natural springs?

JH: It is one of the best ways to see wildlife! True and funny story- I got my husband out on an SUP out in Ft. DeSoto and two manatees swam right up to his board. He did fall in and they swam away, but what a scene to witness! It's very cool what you can see on your SUP out in the open water. I have seen all kinds of fish, manatees, stingrays and birds. It is so peaceful floating around the mangroves and seeing all this wildlife. If you have a waterproof camera, you can get some amazing pictures too! When you are out on the water you really feel like you are one with nature. The fish are right at your fingertips and it really is a cool scene to witness and be a part of.

C: I recently heard about the increasing popularity of using the SUP as a platform for yoga exercises. This is especially interesting to those in the CF community who already practice restorative yoga or one of the many other forms of yoga.

JH: Yoga is all about breathing and balance. I have not tried yoga on a SUP, but I would imagine it could be a good experience. You can put your board on dry land, shallow water or deep water depending on your yoga skill level. Balancing on the board coupled with yoga techniques would prove to be a really high powered yoga session! Yoga is also about mind, body and spirit and what better way to incorporate all of that than out on the open waters with the wildlife and elements surrounding you!

C: Any words of encouragement you wish to share with the CF community?

JH: If you have an opportunity to try SUPing I highly recommend it! It is fun, easy and good for our lungs! ▲

Jennifer is 40 and has CF. She is a Director of USACFA. Her contact information is on page 2. Chris Fowler also is 40 and has CF. He lives in Florida and is an outdoor enthusiast.

In Memory

Debbie Ajini, 42
Shelby Township, MI
Died October 26, 2012

Keven Thomas Kent, 43
South Beloit, IL
Died October 12, 2012

Maureen Lynn Pecor, 48
Melbourne Beach, FL
Died November 2, 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.

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

WELLNESS

Thoughts from a Senior Cystic

By Julie Desch

Hi. I'm Julie and I am a Senior Cystic.

It's time for me to admit and accept this truism.

In standard human years, I am 52. However, in CF years, this equates to 208. Until now, I abhorred the word "cystic". As a noun, I mean. In medicine, behind our backs in the medical school classrooms, nursing stations and hospital hallways, we are referred to as "cystics". I don't know if you knew this or not, but it is true, and it used to make me mad. At least use the whole name of the freakin disease, I thought. But med-speak is truncated, and you and I are cystics. I've decided I like the term now, only because "senior cystic" has such a nice ring to it.

Ironically, I type this as I am prepping for my very first colonoscopy. Somehow, I managed to escape this necessary rite of passage for two years before the Kaiser police found me and reminded me that, at the half-century mark, all of their members are treated to this joyous event.

So the flavored Go-lytely is prepared, the alarmingly large dose of Ducolax swallowed, and my iPhone alarm is set to chime every 15 minutes to remind me to drink up. What better time to write an article about aging with cystic fibrosis?

I had a lovely talk with Dr. Paul Quinton, another Senior Cystic, while in Orlando at the North American Cystic Fibrosis Conference just two months ago. This amazing researcher not only ascertained that chloride transport was messed up (technical term) in CF, he also has managed to live to be approaching 70, and has no plans to end his longevity streak any time soon. That's right… seventy years of age. When describing to me what it felt like to reach his age, he remarked that he was undergoing "total organismal apoptosis". The scientists among you are probably snorting your coffee right now, as I did. It rings so true…so sadly true.

Apoptosis, for those of you who are not cell biology nerds, is the term for programmed cell death. Each of the cells in your body must die at some point, and be replaced by newer, healthy cells to keep your body working. Of course, this is done in a beautifully synchronized way, so we never notice. Parts don't fall off in the normal course of events. Mostly we stay the same, but we get grayer, more wrinkled and blotchy, and slightly more forgetful as newborn baby cells slowly start losing their race with dying cells.

JULIE DESCH, MD

Time to drink.
Disgusting. Did they really think the lemon flavor would disguise anything?

Anyway, back to total organismal apoptosis. I could definitely relate to Dr. Quinton's assessment of the aging of a senior cystic. Many of the changes I've noted are relatively trivial, and can be dispensed with easily. Others are serious, and it is hard to be flippant about them. I'll try to balance these out so as not to write a total downer article. You see, as it turns out, more and more of us are going to get to experience the following, er…, issues. This is the good news. Research and medicine are going to win this war, mark my words. Get ready to age.

Ringing ears: This comes to mind first because, well, my noisy ears are always at the forefront of my mind. It's impossible for that not to be the case. After years and years of tobramycin and similar inner ear poisons, the noise is becoming deafening. I've tried to run away from it (nope), cover my head (nope), talk over it (nope), or drown it out with the TV or music (nope). I've determined that acceptance is the only way to deal. Now I just listen to it when I'm bored, and ignore it when I'm not.

Vision: Going, going, going, gone. I've always been pretty blind, and at the age of 38, had lasik eye surgery thinking that this would hold me over until I was six feet under. A very short time later I needed reading glasses, followed closely by night vision loss. Then even the distance vision that Lasik initially fixed went by the wayside. I gave in a year ago and got progressive lenses. That helped with seeing things either very far away, or very close. Middle distance? Forget it. And don't get me

started about those hair-like floaters that swim around in the periphery.
I hate that alarm.

Hemorrhoids: OK, this is pretty embarrassing, but after years and years of coughing and pooping, this little issue cannot be avoided. It is sort of like the national debt. It just grows and grows as we dilly-dally around the edges with closing tax loopholes (think Tucks pads and Preparation H). We get a bit more serious with "entitlement" reforms—another word I hate—(think banding and injections). But eventually the real knife will be needed. Tax rates will go up and I will be buying Depends.
Why did I ever agree to this procedure?

Complete absence of estrogen: Not a single molecule. Men, you can probably substitute testosterone for estrogen here. We've all probably been a bit low on these sex hormones, especially those of us with a history of questionable nutritional status. But when you add menopause (or andropause) to the already low sex hormone levels…well, it's just not fair now, is it?

I could write a whole article about menopause and CF, but I'll spare you. Hot flashes suck. Tempers flare. Energy takes a nosedive. Sleep? What is that?

Some take exogenous estrogen or testosterone. I choose not to, because I have concerns about estrogen effects on chloride transport. So I deal with hot flashes the same way my mother did: I randomly throw off pieces of clothing and run outside. For sleep, I've tried melatonin (nope), chamomile tea (nope), yoga before bed (nope), and journaling to offload my monkey mind (definitely nope). Ambien works…but it scares me when I begin literally requiring it in order to sleep. So I stare at the ceiling and listen to my ears.

Dementia: You laugh now. Just wait. Names elude me. Sometimes common words elude me in the middle of a sentence. I forget why I walk into a room. I drive by exits. I recite the names of all past dogs before I find the correct one.
Why is that alarm going off again?

Increased Medical Needs: It used to be that all I had to do to take care of myself was take enzymes (and I was horrible at doing that--go figure.) Now, as you probably can relate, this living with CF business is a full-time job. Then, I have to add just normal self-help kind of stuff like daily meditation and exercising and taking wonder supplements and deep breathing exercises (ha), drumming and bingo nights, and well…it is well past the 8:30 pm bedtime set in order to get my necessary beauty rest.

I literally need to make a list of things I need to do in order to remember them all (see dementia above). When on home IVs, I've been known to use a large white board in my office to keep track of everything. Online calendars don't work because I forget to check them (see dementia above).
I can't believe how much of this stuff is left! Bottoms up!

Diminished Lung Function: Hey this happens to everyone with aging, not just us folks with CF. But, of course, it is worse for us. No matter what you do, this happens. So, what is a senior cystic to do?

Tip #1: I keep moving, but I am considering slowing down a bit. I don't mean that I stay in constant motion of course, but every single day I do something active. "Run" is a word that I no longer use. I might jog for short periods of time now, with lovely walking interludes that are marked by impressive bouts of airway clearance. I still lift weights religiously, but I have finally given up on my dream of Olympic Weightlifting competition. My real secret trick is my dogs. They make me move every single day. They must be walked or they drive me crazy, bringing me their leashes, running to the door, bringing my coat to me, tying the poop bag to their collars…impressive given the lack of opposable thumbs.

Tip #2: I rest more. I take frequent naps. I go to bed earlier (so I can stare and listen.) I rest longer between sets when I'm working out. Rest is good. I highly recommend it. The best moment of many days is when I crawl under the covers surrounded by my dogs, knowing that there is nothing I must do for at least eight hours. OK, maybe that sounds a bit pathetic, but it really is a good feeling.

Tip #3: Don't be proud…use supplemental oxygen if you need to in order to keep moving. I haven't had to do this yet. But when I do need to, I will. Guaranteed.
What is that gurgling sound?

Other Organ Issues: Cystic fibrosis is tough on the innards of the body and with age, this becomes quite apparent. The pancreas can totally poop out, and CFRD can result. As of yet, this hasn't happened to me, but as you all know the incidence of CFRD increases with age, so I'm crossing my fingers and toes. I cope by procrastinating my oral glucose tolerance test. The kidneys can get so mad about all of the antibiotics they have filtered that they start to poop out. The liver can scar up, and bile might not flow as well (did it ever?). OK, I'm depressing myself, so I'm going to stop now.
Already? Cheers.

Falling: I am only mentioning this because my dog tripped me while we were running – I mean jogging – yesterday and I have a whopping ankle sprain as a result of falling flat on my face on the sidewalk of a very busy street. I marveled as I hobbled the mile home at how I hadn't broken any bones. I have osteoporosis, of course. As the female senior cystic that I am, it is inevitable. So what's a

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IN THE SPOTLIGHT

With Stephanie Rath

By Andrea Eisenman with Jeanie Hanley

One of the great things about this column, "In The Spotlight", is getting to know individuals one would not normally meet or learn about. For instance, Stephanie Rath. She contacted us to be interviewed and I am so glad that she did. Her verve and outlook on life is humorous and reminded me of the more important things - doing what one can to stay healthy while living one's life and not letting things get her down. And she has had to contend with a lot. Not only daily living with CF but surviving cancer, being on the transplant waiting list for a second time, knowing her limitations and retiring early and above it all, keeping positive. After all, she is married to a handsome, supportive husband plus never gives up doing what she loves–yoga, travel, music and a certain flying animal. Since we did this interview, Stephanie has been elected a Director of USACFA. Congratulations, Stephanie. We are happy to have you with us.

Please welcome our latest star. Spotlight please!

Age: 43

Diagnosed at what age: 19

How: When genetic testing became available in 1989. Until then all my sweat chloride tests were just outside the diagnosable range. Thankfully my doctor always suspected CF and treated me in much the same way.

Once you were diagnosed, what was your reaction to disease?

Well I was 19 and invincible. So I really didn't think that much about it in terms of mortality.

Do you know your CF mutations?

Yes, I'm a Delta F508. I'm really happy for those with G551D right now, though. It sounds like the new Vertex drug Kalydeco will be a big benefit to their health.

STEPHANIE RATH ON A SCOOTER AT MARDI GRAS.

Married? We've been married 15 years and together for 24.

Tell us about your spouse: Randy & I met in college. I started talking to him because he was handsome and was dancing and smiling. Thankfully, he is also a wonderful person. He's been so supportive during all we've been through. He's my best friend. By the way, he's still handsome and still likes to smile and have fun.

Children/do you want them?

We decided early on that it would be too much for both of us to handle considering my health issues.

Occupation/career? I "retired" three years ago on disability. Before retirement I worked at Ernst & Young LLP as a tax manager in their real estate practice. The work was very challenging and I enjoyed the intellectual stimulation and relationships. Unfortunately, the busy seasons were just too much for me. However, the firm has been so great at accommodating me and has excellent benefits. I'm very lucky.

Who or what made you get Long Term Disability (LTD) benefits from your former employer?

When I started to work there my benefits package included LTD coverage. Because I opted to pay the premium, my income from it is tax-free. I also had no pre-existing rider because of signing on when hired. For me it was a no-brainer to take the coverage. My past history told me how life with CF can get really scary really quickly. Later a buy-out option became available, which I took. I'm so incredibly thankful to be so lucky.

Have you considered a lung transplant?

Yes, twice. Back in 1992 (age 23) I developed a bad pneumonia, was put on 3L of O_2 24/7, and was listed for a double lung transplant. It was a big shock to me at the time. I had been on IVs before and even in the hospital. But I'd never really felt like I was going to die. It was a huge wake up call.

In 1992 pulmonary rehab was not available. So the transplant program had you meet with a physical therapist once and they gave you a few suggested exercises. I started using an aerodyne bike we had at home. At first, three minutes was all I could do. Every day I tried to add a minute if I could. I REALLY wanted to live. Being in love with Randy was great motivation. Eventually I was able to get off oxygen, return to work full time, and was delisted for transplant. However, significant damage had been done and my FEV_1 stabilized at around 40%.

Now almost twenty (yes, that's right - twenty) years later I'm consid-

ering a first transplant again. My FEV_1 has dropped to 26%. Now I need oxygen at night and sometimes with excessive exertion. My exacerbations have gotten more frequent and my antibiotic sensitivity is pretty poor.

Duke Medical evaluated me in August 2010. Right after the workup testing, I decided to have a colonoscopy. I was having blood in my stool and thought it should be checked out. Anticipating hemorrhoids, a diagnosis of rectal cancer was a real shocker. One fatal disease I can handle – but two? REALLY??? So began a course of treatment including chemo and radiation and a colostomy surgery. Thankfully, my lymph nodes were clean.

Most transplant centers have a rule that you must be cancer free for two years before transplant. Thankfully, I only have about eight months left, until December 2012, before I can be listed. If you're considering a transplant start the testing sooner rather than later. Then if you have any other issues that pop up they can be addressed early on.

What are your fears about transplantation?

To be honest I'm not really afraid of the surgery. My biggest concern is post-transplant longevity. But the reality is, when it's time for me to be listed, I will feel like I'm waiting for the best Christmas present ever.

What made you interested in yoga?

My uncle John gave me a yoga DVD when I was sick once. It made me feel so at peace that I was hooked. It also really helped me learn to control my breath in a calm way. John also took me to my first class with Nikki Myers. Just before my cancer diagnosis I completed my 200-hour teacher certification at her studio. The training was my way of deepening my own practice. However, I would love to volunteer as an instructor for people with pulmonary rehab needs.

Do you think you would be alive today without exercise?

Absolutely not! Exercise has helped me keep my lungs open and strengthen both my body and immune system. If I take more than a few days off I really feel it. It is the single most important adjunctive therapy I know of.

What are you able to do now?

There are good days and bad days. On good days I spend time with friends, do household errands and exercise. On bad days I take a nap and try to do things that take less energy like paying bills. I always try to make it to pulmonary rehab. I'm lucky to have more time to spend with my mom during my retirement. Retirement has also given me time to learn new things like sewing.

What do you do for fun?

We really love to travel. Many of our trips involve music and/or camping. With oxygen and medical equipment, that becomes a bit more complicated. But I'm not about to be limited by the oxygen any more than is necessary. One of our trips we camped in Black Rock Desert in Nevada. We go on a music cruise every year and have made friends all over the country on it. We visited one of them on our recent trip to Mardi Gras. Our friend showed us the locals' version, which was amazing. Thankfully, I had a scooter lent to me by another friend with CF who is post-transplant. It was great to be able to keep up with everyone else using my non-"hoverround" scooter.

What is your favorite music?

I have pretty diverse tastes including rock, bluegrass, funk, disco, jazz, jam-bands, reggae, blues, etc. Funk and disco are really great for hula hooping.

Discuss what having dogs has been like for you:

My Boston Terriers, Rocky & Rosie, are the best. They remind me to be happy every day. They are also great snugglers and have a profound sense of when you're not feeling well.

What made you want to do a blog?

I never envisioned being a "blogger". But the more I got sick the more people wanted to know how I was doing. It became a way for me to keep friends and extended family up to date on my health.

What is your blog about?

Primarily it's about my experiences with my health issues. It's part medical update and part journal. It helps me deal with some of the emotions that come along with this disease. I also cope and get inspired by reading others blogs. The CF community is such a great source of support especially when dealing with big issues like transplant. Blogging is a great way of documenting shared experiences. My blog is: www.ifpigshadwings.com.

Tell us about your love of Flying Pigs and why?

As a kid I always loved pigs. Charlotte's Web was one of my favorite books. One time my Grandma gave me a small figure of a pig with wings. That's when the love of flying pigs began. For me a flying pig is a symbol of overcoming the impossible. Looking at a cute pig with wings, flying and smiling, really motivates me to do the same every day.

My blog web address is inspired by comedian Heywood Banks' song "If Pigs Had Wings". The chorus is "If pigs had wings, away they'd fly up into heaven oh so high. If pigs had wings away they'd soar up in through heaven's golden door." When things get really tough, this song reminds me that I will be OK no matter what.

What do you look forward to in life?

I look forward to not only living as long as I can but with as many adventures as I can.

Are you an optimist?

I'm definitely a glass half full per-

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UNOS LAS System Needs Tune-Up While Major Lack Of Organ Donors Still Main Problem

By Steve Jenkins

It is really an unfortunate circumstance people find themselves in when they live in a world and country where you have to wait an unnecessarily long time to get an organ transplant. It's not right that people who need new organs are made to suffer for unnecessarily long times. The two reasons for this are: the United Network of Organ Sharing (UNOS) use of the Lung Allocation System (LAS) and a national shortage of organ donors.

UNOS is the governing body that created the new points system to allocate organs. The LAS for pre-transplant patients uses a points and list system where the sicker you are and the worse your health is, the sooner you get a transplant. The LAS system was an improvement from the previous system which used time waited on the list as the only factor. The positive side of the new LAS is more people's lives are being saved. That is great and I applaud UNOS for doing this. But there is also a negative to the LAS system. It causes worse post-transplant outcomes. It is likely this is because of a national and worldwide shortage of organs, since more people aren't organ donors. Unfortunately, some people decline to be organ donors because of lies and misconceptions they have about organ donation. The system needs fixing, no doubt. But the main problem is a lack of organ donors.

I must, as it is necessary, acknowledge that lungs can often be difficult to harvest. That is a problem that needs to be addressed with more research into better harvesting and procurement techniques, which would yield more organs for transplantation. But still, the main problem is there are not enough organ donors out there.

The reason for the lack of donors is that people are inherently fearful. We are easily scared and put into a state of fear. For example, the potential threat of terrorism has many in this country living in fear. This applies to organ donation in that people are fearful that doctors will let them die on purpose in order to take their organs. In fact, this is a complete and utter falsehood. However, it is a widely believed urban myth and makes people who believe it opt out of being an organ donor. After all, once you have passed away, your body is no longer alive. It is nothing more than molecules and atoms and is going to waste away. So why not allow as much of your body as possible to be given to others to save their lives? What better way to go out of this world, than by saving the lives of others through your generosity and selflessness? It only makes sense to me to donate not only your organs when you pass but also your body for research to advance future medicine.

Some people aren't organ donors because they are fearful. There are likely other reasons people do not become organ donors, but I believe fear is the main issue concerning the organ donation shortage. One remedy for this problem is the use of an opt-out system, which is currently used in many countries in Europe. The opt-out system automatically makes everyone an organ donor, unless you opt out. It yields many more organ donors. The current system in the United States is an opt-in system, where you must sign up to be an organ donor. The United States should change to the opt-out system.

Personally, I am a recipient of two lungs. I needed new lungs because I have cystic fibrosis (CF), which caused my lungs and airways to be clogged with mucous and infection and to function at only 17%. My donor was a 23-year-old young man who was struck by a speeding car while crossing the street. He was deemed brain dead sometime after arriving at the hospital. Not only did he donate two lungs to me, he donated his heart, kidneys and liver as well. My donor was a committed organ donor and his family supported his decision. His passing was a tragedy but much good was able to come from it. That's what life is all about, making something good out of something bad. That's what organ transplantation is all about, making something good out of something bad. From the unfortunate passing of a person arose the creation of a miracle in the saving of five other people's lives.

My donor went out of this world as a hero for having saved the lives of five people. Society should revere organ donors who save the lives of others. Those who have passed and donated their organs should be thought of as heroes. Those who have not yet passed but are registered organ donors should be viewed with high esteem. Not too many things are more heroic than saving the lives of others.

I am forever grateful for my donor, his family and loved ones and the

STEVE JENKINS, CENTER, WITH HIS COUSIN CORY HARTZE, TO THE LEFT, AND SISTER, KELSEY JENKINS.

miracle that happened as a result of their tragedy. I cannot imagine what it is like to lose a close family member but hope they gain some peace in knowing their tragedy resulted in five miracles.

I am an organ and tissue donor. Many of my family and friends are organ donors due to my situation. In an ideal world, every person would be able to say that about their family and friends. Those of us who believe in organ donation need to do a better job and work harder on spreading organ donation awareness and truth. The fear of what might happen to you if you are an organ donor and in critical care in the hospital doesn't have to exist. With better education and awareness about the current organ donation system and the need for more organs, people will have their organ donation fears alleviated. They would no longer live in the fear of doctors letting them pass in favor of harvesting their organs. If that happened, it is logical many more people would become organ donors.

According to DonateLife, the leading advocacy group for organ donation, in 2008 only 38.2% of people, or 79.7 million people were registered organ donors across all 50 states in the United States of America.

If more people became organ donors, it would increase the number of organ transplants. This would lead to two things: more lives saved and ultimately better transplant outcomes. First, more lives would be saved because, with no shortage of available organs, anyone who needed a transplant would be able to get one without waiting very long. This means more people would make it to transplant, which would lead to more lives being saved. Second, and just as important, is better transplant outcomes. Since the new UNOS Lung Allocation System was put into place in 2005, lung transplant success rates have been down. This is due to the fact that people have to get very sick before they get their needed lungs. The sicker you are going into a transplant, the worse you will do after a transplant. The healthier you are going into a transplant, the better you will do after a transplant. Shouldn't the system be implemented in such a way that this is part of its main focus? The current UNOS system was not implemented with this as its main focus. Instead, it was implemented with the main goal being saving more lives.

Indeed, the current UNOS system has saved more lives than the previous system. But in doing this, they have created a system where people are usually only transplanted when they are very sick, which leads to worse outcomes post-transplant. This is fair to one group of patients and unfair to another group. It is fair to those whose health declines rapidly and need an organ transplant or they will die very soon. For them, the system works well because it saves their lives. They would otherwise have died had they not gotten the transplant.

Pre-transplant patients hope to live long lives post-transplant. The new UNOS Lung Allocation System is unfair to those patients who are a bit healthier and want the transplant as soon as possible, mainly because they know that the healthier they are going into transplant the better their post-transplant outcome. I fall into this category. Let me point out that I am on the list for my second double-lung transplant, and I fully agree and respect that anyone going for a first transplant should get a transplant ahead of me. It's only fair. What isn't fair is that there is a potentially infinite number of times I could be passed over by other patients on the waiting list who are sicker than me. This causes me to have to wait unnecessarily long and continue to live a very poor life.

My health hasn't declined for more than two years and my lungs are currently functioning at 22%. How

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KELLEY *continued from page 13*

lism and bone. Thus, kidney function should also be evaluated, particularly in the evaluation of decreased BMD. A lateral spine film should be considered in an individual with decreased BMD to assess for presence of vertebral compression fractures as these can be asymptomatic.

Prevention of poor bone health in CF focuses on overall good nutrition, adequate supplementation of calcium, vitamin D (goal 25-hydroxy vitamin D level > 30 ng/mL) [>75 nmol/L; 1.0 nmol/L = 0.4 ng/mL], and other vitamins and minerals, regular exercise, and avoiding excessive glucocorticoid use if possible.

Treatment is indicated for BMD-Z/T <-2. For individual's whose DXA BMD-Z/T is between -1 and -2, treatment should be considered if the individual is being listed for transplant, has had what is referred to as a fragility fracture (spine, rib), or if excessive loss has occurred. How excessive/accelerated is defined is not clear—in children, though, bone density should be increasing and certainly not decreasing.

Treatment options for osteoporosis in the elderly have expanded over the past few years. The extent to which these newer therapies will be effective and safe in CF has not yet been determined. Current recommendations are to confirm optimal vitamin D status and adequate calcium, zinc, and vitamin K intake. Bisphosphonates are the mainstay of therapy in CF. These medications prevent bone loss. Pamidronate and zoledronate (both delivered intravenously) and aledronate (an oral form) increase bone mineral density and appear to decrease fracture risk in adults. In general, limited data regarding bisphosphonate use in children, including those with cystic fibrosis, is available (this group of medications has been used in children with a bone disorder known as osteogenesis imperfecta).

In the otherwise healthy population, increasing treatment strategies are becoming available to prevent and treat osteoporosis in the elderly. Attention, including commercial ad campaigns, is also focusing on prevention of osteoporosis in the older population by targeting childhood and adolescence, the origin of peak bone mass. Given the risk of poor bone health in younger adults with cystic fibrosis, these same strategies of prevention, screening, and exploration of treatment options that minimize fracture risk and optimize life quality are crucial. As individuals with cystic fibrosis age and postmenopausal and senile osteoporosis are additional realities, the role of bone preservation in childhood and early adult years becomes even more relevant. ▲

Dr. Kelley is an Attending Physician, Division of Endocrinology & Diabetes and Assistant Professor of Pediatrics, Perelman School of Medicine at University of Pennsylvania.

TILLMAN *continued from page 20*

Cystic Fibrosis(NACF) Conference, Orlando, Florida, October 11-13, 2012

The first phase II study combining CFTR-targeted therapies reports clinically meaningful improvements in lung function in F508del homozygous CF patients. VX-809, now called lumacaftor, is a CFTR corrector that improves CFTR trafficking.
http://tinyurl.com/budaf7d

SINUS

Colonisation and infection of the paranasal sinuses in cystic fibrosis patients is accompanied by a reduced PMN response. Helle Krogh Johansen, Kasper Aanaes, Tania Pressler, Kim Gjerrum Nielsen, Jacob Fisker, Marianne Skov, Niels Høiby, Christian von Buchwald. Journal of Cystic Fibrosis. Volume 11, Issue 6 , Pages 525-531, December 2012

The authors studied whether the sinuses might be a principal site for *Pseudomonas aeruginosa* lung infection. They have shown that *P. aeruginosa* form biofilm in the sinuses, which constitute an important bacterial reservoir for subsequent lung infection. The high amount of IgA in the upper airways probably protects *P. aeruginosa* from the inflammatory immune system, and they can proceed unnoticed into a permanent infectious site that cannot be eradicated with antibiotics.
http://tinyurl.com/agwdedb

INFECTIONS/EXACERBATIONS/TREATMENTS

Respiratory virus infections commonly associated with exacerbations. Report From the 26th North American Cystic Fibrosis (NACF) Conference, Orlando, Florida, October 11-13, 2012

A one-year observational study in Manchester, UK, reports that respiratory virus infections are common among adult patients with CF, and that infections are commonly associated with pulmonary exacerbations. The most common viral pathogens were rhinovirus, metapneumovirus, adenovirus, and influenza A. Viral infection was associated with a two-fold higher risk of an exacerbation compared to no viral infection. Pulmonary exacerbations, but not viral infection, were associated with a decline in FEV_1 in this study. A separate study in the US has used live-cell imaging technique to show that RSV infection promotes the growth of biofilm by *Pseudomonas aeruginosa* (Fischer et al. NACF 2012; abstract 262), suggesting one way that viral co-infection may worsen lung function in CF. A similar effect on *P. aeruginosa* biofilm

RATH *continued from page 27*

son, most of the time. Sometimes the glass size gets bigger, though, and you need to find a way to fill it back up. That's when I call on the flying pigs to haul buckets of water.

Final thoughts?

Try to do your best to stay as healthy as you can as long as you can. Then you've done all that you can do and there is no regret. Also don't forget to have fun along the way. ▲

"Life is as easy or as hard as you think it is."

– Jonathan Lockwood Huie

Andrea Eisenman is 48 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. Jeanie Hanley is 50 and is a physician who has CF. She is a Director of USACFA and is the Vice President. Her contact information is on page 2. If you would like to be interviewed for In The Spotlight, contact either Andrea or Jeanie.

DESCH *continued from page 25*

girl to do? My recommendation is to avoid falling at all costs, and to lift weights, take vitamin D, magnesium, drink your milk, and pay close attention to your dog if a squirrel should appear in her line of sight.

Shrinking: This sounds odd, but it is literally true. Even "normal" people begin to lose muscle mass after the age of 30. I've read of different rates, but basically the older you get, the faster muscle wastes away. But, this does not have to be the case! Just as healthy people can moderate this muscle loss by appropriate weight lifting exercises done consistently, so can we.

We also tend to shrink vertically more than the average bear. "Normal" people get shorter as their intervertebral disks compress with age. That happens to us, too; but in addition to that, we tend to hunch forward more due to weird mechanical changes in our thoracic spines. The result: more vertical shrinkage.

There is not a lot to do about this. I work on my T-spine mobility every day, trying to prevent the hunchback thing. I also just ordered an inversion table so I can hang upside down. I may combine that with some airway clearance. We'll see.

Uh oh…good thing I'm using a laptop.

Loss: This is the hardest of all to cope with. We lose our friends and family members as they succumb to this awful disease before we do. There is no making light of this. It's not just sadness and grief that we live with, but for me, there is also guilt. Why do I get to live so long and enjoy family and friends when my loved ones and close friends suffer and die? Life is not fair, and there is no joke to soften the blow of this fact. ▲

Julie is 52 and is a physician who has CF. She may be contacted at: jdesch@usacfa.org.

TILLMAN *continued from page 30*

was seen when airway cells were co-infected with rhinovirus and adenovirus. Viral infections appear to induce a more rapid maturation of biofilm.
http://tinyurl.com/ch8e6se

Iron chelation: effects on *P. aeruginosa* biofilm. Report From the 26th North American Cystic Fibrosis (NACF) Conference, Orlando, Florida, October 11-13, 2012

Recent laboratory studies have indicated that iron plays an important role in P. aeruginosa biofilm formation in the CF lung. Airway cells expressing F508del release more iron than cells rescued by wild-type CFTR, and this iron promotes growth of P. aeruginosa and enhances biofilm formation. A follow-up study in CF airway epithelial cells found that adding an iron chelation agent (deferoxamine or deferasirox) to tobramycin reduced biofilm by about 90% and reduced bacterial density by 7-log units.
http://tinyurl.com/cr3bo3j

Treatment of lung infection in patients with cystic fibrosis: Current and future strategies. Döring G, Flume P, Heijerman H, Elborn JS; for the Consensus Study Group. Journal of Cystic Fibrosis. 2012 Dec;11(6):461-479.

In patients with cystic fibrosis lung damage secondary to chronic infection is the main cause of death. Treatment of lung disease to reduce the impact of infection, inflammation and subsequent lung injury is therefore of major importance. Here the present status of antibiotic therapy for the major pathogens in CF airways is discussed, including prophylaxis against infection, eradication of early infection, suppression of chronic infection, and the treatment of infective exacerbations. Measures to optimize maintenance treatment for infection are outlined in the light of novel antibiotic drug formulations. New developments in culture-independent microbiological diagnostic techniques and the use of tools for monitoring the success of antibiotic treatment courses are discussed. Finally, cost-effectiveness analyses for antibiotic treatment in CF patients are discussed.
http://tinyurl.com/cf9ppqj

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. Volume 11, Issue 5, Pages 458-460

The authors report the case of a

Continued on page 32

JENKINS *continued from page 29*

hard is it to live and breathe? Try sticking a coffee stirrer in your mouth, closing your lips and plugging your nose and breathing. See how hard it is to breathe? That is as hard as it is to take each and every breath right now. All day, every day. Walking two blocks feels like I just sprinted a mile. Anyone who has waited for a lung transplant knows exactly what this feels like. It is not a life I wish upon anyone. I am made to live with this situation for an unnecessarily long time because of the way the UNOS Lung Allocation System works. Just as bad is that I must suffer even more and get even sicker before I will get re-transplanted. This could potentially lead to a worse success rate of my re-transplant.

This frustrates me very much. Why should a system be able to potentially control the success rate of my future transplant? Lower success rates are justified by stating that more lives have been saved.

The UNOS Lung Allocation System needs serious review. Success rates of transplants are down and that is not good news for the transplant community. UNOS has forced the medical system to operate in a way that compromises life expectancy of people post-transplant. The UNOS system is unfair to people whose health doesn't rapidly decline and who are made to wait longer and possibly live shorter lives. Something needs to be done about this.

An improvement of the UNOS system would greatly benefit transplant recipients. If the right corrections were made, it would make it possible to get transplanted without being extremely sick. It could make people have better transplant outcomes. It would also decrease the amount of waiting time pre-transplant patients have to go through.

However, much bigger than the UNOS Lung Allocation System problem is the organ donation problem. If more people were less fearful, then more people would become organ donors. This would lead to two things. First, many more lives would be saved because everyone who needed an organ or organs would get them. Second, transplant outcomes would be much better because people who were healthier would be getting transplanted sooner. Sooner transplants lead to better outcomes.

Currently, only 26 states offer the driver's license option to become an organ donor. This method is simple and requires nothing more than a signature next time you get your license renewed. Even better and more efficient, you can become an organ donor now by simply visiting DonateLife's FaceBook page at www.facebook.com/donatelife and click on the "Register" tab. ▲

Steve is 29 and has CF. He had a bilateral lung transplant on March 6, 2007 and is on the waiting list for a re- transplant. He lives in Kent, WA.

TILLMAN *continued from page 31*

38-year-old man, known to have chronic infection with the ET12 strain of Burkholderia cenocepacia who developed cepacia syndrome 26-years after initial infection. Aggressive treatment with a combination of 4 intravenous antibiotics, oral corticosteroids and cyclosporin brought about clinical, radiological and biochemical resolution of his cepacia syndrome. This case highlights the possible role of cyclosporin in the treatment of cepacia syndrome.
http://tinyurl.com/8r7xauj

Inhaled aztreonam lysine vs. inhaled tobramycin in cystic fibrosis: A comparative efficacy trial. Baroukh M. Assael, Tacjana Pressler, Diana Bilton, Michael Fayon, Rainald Fischer, Raphael Chiron, Mario LaRosa, Christiane Knoop, Noel McElvaney, Sandra A. Lewis, Mark Bresnik, A. Bruce Montgomery, Christopher M. Oermann. Journal of Cystic Fibrosis. Published online 17 September 2012.

An open-label, parallel-group, international trial comparing aztreonam for inhalation solution (AZLI) and tobramycin nebulizer solution (TNS) for cystic fibrosis patients with airway *Pseudomonas aeruginosa* was conducted. AZLI demonstrated statistical superiority in lung function and a reduction in acute pulmonary exacerbations compared to TNS over 3 treatment courses.
http://tinyurl.com/cwval2n

A network meta-analysis of the efficacy of inhaled antibiotics for chronic Pseudomonas infections in cystic fibrosis. Kavi J. Littlewood, Kyoko Higashi, Jeroen P. Jansen, Gorana Capkun-Niggli, Maria-Magdalena Balp, Gerd Doering, Harm A.W.M. Tiddens, Gerhild Angyalosi. Journal of Cystic Fibrosis. Volume 11, Issue 5, Pages 419-426, September 2012.

Various inhaled antibiotics are currently used for treating chronic *Pseudomonas aeruginosa* lung infection in cystic fibrosis (CF) patients, however their relative efficacies are unclear. The efficacy of the inhaled antibiotics tobramycin (TIP, TIS-T, TIS-B), colistimethate sodium (colistin) and aztreonam lysine for inhalation (AZLI) based on data from randomised controlled trials was studied. The authors conclude that all studied antibiotics have comparable efficacies for the treatment of chronic P. aeruginosa lung infection in cystic fibrosis (CF).
http://tinyurl.com/9zbvlqh

Identification and distribution of Achromobacter species in cystic

fibrosis. Theodore Spilker, Peter Vandamme, John J. LiPuma. Journal of Cystic Fibrosis. Published online 08 November 2012.

The use of *nrdA* sequence analysis allows differentiation of the several *Achromobacter* species that can infect persons with CF. *Achromobacter* species other than *A. xylosoxidans* account for the majority of *Achromobacter* infection in CF patients in the U.S.
http://tinyurl.com/d6jtgxg

Extending the course of intravenous antibiotics in adult patients with cystic fibrosis with acute pulmonary exacerbations. Sequeiros, IM, Jarad, NA. Chronic Respiratory Disease, 11/09/2012. vol. 9 no. 4 213-220

Most severe pulmonary exacerbations (PExs) in adult patients with cystic fibrosis (CF) are treated with 2 weeks of intravenous (IV) antibiotics. At occasions, the treatment is extended. PExs in patients with worse lung disease and greater residual symptoms and lung inflammation at day 14 of antibiotic treatment were associated with the extension of the course of IV antibiotics. Prolonging the treatment to 21 days improved symptoms, but did not result in improvement in any other short–term or lung outcome measures.
http://tinyurl.com/avu99lv

Safety, efficacy and convenience of colistimethate sodium dry powder for inhalation (Colobreathe DPI) in patients with cystic fibrosis: a randomised study. Antje Schuster, Cynthia Haliburn, Gerd Döring, Martin Harris Goldman. Thorax. Published online 11/7/2012

The study aims to assess efficacy and safety of a new dry powder formulation of inhaled colistimethate sodium in patients with cystic fibrosis aged ≥6years with chronic Pseudomonas aeruginosa lung infection (CDPI). CDPI demonstrated efficacy by virtue of non–inferiority to tobramycin inhaler solution in lung function after 24weeks of treatment. There was no emergence of resistance of P aeruginosa to colistin. Overall, CDPI was well tolerated.
http://tinyurl.com/at6bnux

Fluoroquinolones in the treatment of bronchopulmonary disease in cystic fibrosis. Matthew Hurley, Alan Smyth. Therapeutic Advances in Respiratory Disease. December 2012 vol. 6 no. 6 363-373

Fluoroquinolones are commonly used to treat lung infections in patients with cystic fibrosis. These patients are susceptible to lung infection with common bacteria such as Staphylococcus aureus and Haemophilus influenzae, but are also prone to infection by opportunistic bacteria, including Pseudomonas aeruginosa. The good oral bioavailability and broad antimicrobial spectrum of activity, including antipseudomonal properties, make this class of antimicrobial attractive.
http://tinyurl.com/blmmu6d

Pooled analysis of two large randomised phase III inhaled mannitol studies in cystic fibrosis. Diana Bilton, Gabriel Bellon, Brett Charlton, Peter Cooper, Kris De Boeck, Patrick A. Flume, Howard G. Fox, Charles G. Gallagher, David E. Geller, Eric G. Haarman, Helge U. Hebestreit, John Kolbe, Allen Lapey, Phil Robinson, Jian Wu, Jonathan B. Zuckerman, Moira L. Aitken. Journal of Cystic Fibrosis. Published online 11 December 2012.

Sustained six-month improvements in lung function and decreased pulmonary exacerbation incidence indicate that inhaled mannitol is an important additional drug in the treatment of CF.
http://tinyurl.com/cx5k6cc

Eradication therapy for *Pseudomonas aeruginosa* colonization episodes in cystic fibrosis patients not chronically colonized by *P. aeruginosa*. Petra Schelstraete, Filomeen Haerynck, Sabine Van daele, Sarah Deseyne, Frans De Baets. Journal of Cystic Fibrosis. Volume 12, Issue 1, Pages 1-8, January 2013

In this article, an overview on the natural history of early *Pa* colonization and the history of eradication treatment is given. Moreover, the results of the different eradication treatment trials and directions for future research are discussed.
http://tinyurl.com/brqekbm

***Mycobacterium avium* and *Mycobacterium abscessus* complex target distinct cystic fibrosis patient subpopulations.** Emilie Catherinot, Anne-Laure Roux, Marie-Anne Vibet, Gil Bellis, Sophie Ravilly, Lydie Lemonnier, Evelyne Le Roux, Claire Bernède-Bauduin, Muriel Le Bourgeois, Jean-Louis Herrmann, Didier Guillemot, Jean-Louis Gaillard. Journal of Cystic Fibrosis. Volume 12, Issue 1 , Pages 74-80, January 2013

Mycobacterium avium complex affects adult patients with a mild form of CF, whereas Mycobacterium abscessus complex affects younger patients with more severe CF and more frequent intravenous antimicrobial treatment.
http://tinyurl.com/cgc7vky

FYI

Cystic Fibrosis: What to Expect now in the Early Adult Years. Bradley S. Quon, Moira L. Aitken. Paediatric Respiratory Reviews. Volume 13, Issue 4, Pages 206-214, December 2012

This article discusses the emergence of chronic disease–related co–morbidities such as CF–related diabetes, chronic kidney disease, bone disease, arthropathy, and depression. It also provides an overview of the many challenges confronted by adult CF care providers.
http://tinyurl.com/b9wncj3

Cystic Fibrosis: Management of Haemoptysis. K. Hurt, N.J. Simmonds. Paediatric Respiratory Reviews.

Continued on page 34

Volume 13, Issue 4, Pages 200-205, December 2012

Haemoptysis is a common complication in cystic fibrosis (CF), occurring in approximately 9% of the population. Massive haemoptysis is associated with older patients, more severe disease and carries a high mortality rate. Despite this there are few robust published studies of effective treatments and knowledge of the precise pathogenesis is limited. Current guidelines for treatment from the Cystic Fibrosis Foundation (CFF) are based on consensus opinion of experts. Patients with massive haemoptysis who do not respond to initial medical treatments should undergo bronchial artery embolization.

http://tinyurl.com/b7w5nt2

Soluble squalamine tablets for the rapid disinfection of home nebulizers of cystic fibrosis patients. Lamia Djouhri-Bouktab, Kamel Alhanout, Véronique Andrieu, Nathalie Stremler, Jean Christophe Dubus, Didier Raoult, Jean Marc Rolain, Jean Michel Brunel. Journal of Cystic Fibrosis. Volume 11, Issue 6, Pages 555-559, December 2012

The bacterial contamination of nebulizers represents a major problem for cystic fibrosis patients that can lead to reduced nebulizer performance and increase the risk of patient reinfection by the contaminating bacteria. The authors investigated the potential use of squalamine, a broad–spectrum antimicrobial compound, as a nebulizer disinfectant. The results suggest that amino sterol derivatives may be used by cystic fibrosis patients for rapid and easy home nebulizer disinfection and that soluble tablets may be developed for this purpose.

http://tinyurl.com/bqmvqye

Cancer Risk in Cystic Fibrosis: A 20-Year Nationwide Study From the United States. Patrick Maisonneuve, Bruce C. Marshall, Emily A. Knapp and Albert B. Lowenfels. Journal of the National Cancer Institute, Published online 11/24/2012

The aim of this study was to determine cancer risks in non-transplanted and transplanted CF patients. The overall burden of cancer in CF patients remains low; however they have an increased risk of digestive tract cancer, particularly following transplantation. They also have increased risk of lymphoid leukemia and testicular cancer, and decreased risk of melanoma.

http://tinyurl.com/csstljk

Nebulized hypertonic saline containing hyaluronic acid improves tolerability in patients with cystic fibrosis and lung disease compared with nebulized hypertonic saline alone: a prospective, randomized, double-blind, controlled study. Maria Lucia Furnari, Lisa Termini, Gabriella Traverso, Stefania Barrale, Maria Rita Bonaccorso, Giuseppina Damiani, Caterina Lo Piparo, Mirella Collura. Therapeutic Advances in Respiratory Disease. December 2012 vol. 6 no. 6 315-322

The authors carried out a prospective, randomized, double-blind, parallel-group, controlled study of a hypertonic saline solution containing hyaluronic acid (Hyaneb) versus standard hypertonic saline therapy to assess whether the presence of hyaluronic acid would improve the tolerability of hypertonic saline. The results showed that nebulized Hyaneb was more effective in reducing the need for bronchodilators and caused a significant reduction in the incidence of adverse effects compared with nebulized hypertonic saline solution alone. Its safety profile indicates that Hyaneb can be used for the treatment of lung disease in cystic fibrosis.

http://tinyurl.com/cvn85b3

Longitudinal Cystic Fibrosis Care. S S Antunovic, M Lukac and D Vujovic Clinical Pharmacology & Therapeutics (2013); 93 1, 86–97.

Cystic fibrosis is a complex disease entity that presents considerable lifelong challenges. Implementation of medical and surgical treatment options involves multisystem interventions to prevent and treat lung and gastrointestinal manifestations of cystic fibrosis and associated comorbidities. From birth through adulthood, cystic fibrosis care entails a longitudinal regimen aimed at achieving relief of disease symptoms and enhanced life expectancy. With increased knowledge of the molecular behavior of the cystic fibrosis transmembrane conductance regulator (CFTR) in health and disease, clinical practice has been enriched by the prospect of novel strategies, including mutation-specific drug and gene therapy targeting restoration of corrupted transepithelial ion transport. Emerging paradigms of comprehensive care increasingly enable personalized solutions to address the root cause of disease-transforming management options for individuals with cystic fibrosis.

http://tinyurl.com/cdmjl57

Recurrent *Clostridium difficile* colitis in cystic fibrosis: An emerging problem. Katarine Egressy, Michaelene Jansen, Keith C. Meyer. Journal of Cystic Fibrosis. Volume 12, Issue 1, Pages 92-96, January 2013

C. *difficile* colitis can cause life threatening illness in patients with CF, and symptoms may be subtle and/or atypical and lead to significant delay in diagnosis. Patients with recurrent C. *difficile* colitis are at high risk of fatal outcome, and empiric therapy should be considered for patients with previous C. *difficile* colitis even in the absence of disease when broad-spectrum antibiotics are given to treat bacterial infection.

http://tinyurl.com/ceggvzo

LUNG TRANSPLANT

Gastroesophageal reflux disease in lung transplant patients with cystic fibrosis. Bernardino M. Mendez, M.D., Christopher S. Davis, M.D., M.P.H., Cynthia Weber, M.D., Raymond

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TILLMAN *continued from page 34*

J. Joehl, M.D., F.A.C.S., P. Marco Fisichella, M.D., F.A.C.S. The American Journal of Surgery. Volume 204, Issue 5, Pages e21-e26, 11/19/2012

The authors compared esophageal pH monitoring, manometry, gastric emptying studies, and barium swallow of 10 lung transplant patients with cystic fibrosis with those of 78 lung transplant patients with other end-stage pulmonary diseases. Lung transplant patients with CF have a significantly higher prevalence and proximal extent of gastroesophageal reflux disease (GERD) than do other lung transplant recipients. These data suggest that CF patients in particular should be routinely screened for GERD after transplantation to identify those who may benefit from antireflux surgery, especially given the risks of GERD-related aspiration and chronic allograft injury.
http://tinyurl.com/c3lzmxy ▲

Laura Tillman is 65 and has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.

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- 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

PRINTED WITH SOY INK

Published by the United States
Adult Cystic Fibrosis Association, Inc.
CF Roundtable is printed on recycled paper.

IMPORTANT RESOURCES

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

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

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

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

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