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

Grandpa Counts His Blessings

By Paul Feld

There are many milestones in life. Some you plan on and count as goals, and others just randomly happen when you least expect it. As someone diagnosed with CF as a teenager in 1976, becoming a grandpa was never, ever a goal in my life. So let's just say that when I became a grandpa on March 26, 2013, it was a random event that I never expected.

A little explanation and history is warranted here. After I was diagnosed, at the age of 19, I was not told that almost all CF males were infertile. I married my first wife when I was 22 and, after a year or so, we decided to try to have children. After another six months, with no success, we spent a lot of money trying to figure out what was wrong with HER. Of course, nothing was found. We then did a relatively simple sperm count test on me, which found a total sperm count of zero. Yes, zero.

Of course, that could not be right. So we did it again - with the same result. Needless to say, our hopes were dashed that we'd have our own child. In-vitro fertilization was in its infancy and the costs were more than prohibitive to us, a newly-married couple.

PAUL FELD AND NEW GRANDCHILD, CHRISTOPHER JAMES.

It quickly appeared our only option was adoption. We worked with Catholic Charities in St. Louis and submitted an application. While both my wife and I had decent employment and we had a starter home, the agency did not consider a couple with a daddy having CF to be the best applicants in the world. They asked my pulmonary physician about how long I could expect to live because, quite frankly, at that time I had already reached the median age for CF survival. My physician promptly responded that he had no idea how long I, or any of his patients, might live. He was a man of faith and believed only God knew the answer to that. It was probably the best response he could have given. After a lengthy 'thinking' period, the agency agreed that we could be parents to a child in their care, but they did state that we could handle only one

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EDITOR'S NOTES

I hope all of you have read the front page article about **Paul Feld** becoming a grandfather. In many groups that would be of no significance but, in our group, it is of great significance. Many of us were diagnosed when people still were being told that their children who had CF would not live to adulthood – let alone get married, have children and live to see them to adulthood, and become grandparents. Hooray for all of us!

Another bit of USACFA news is that **Stephanie Rath** is on leave of absence while she waits for new lungs. We wish her the best and hope for a speedy and uneventful recovery. We'll keep you posted on her.

One more bit of USACFA news is that we are completely free-of-charge now, thanks to all our donors and sustaining sponsors. The founders hoped that the newsletter could be offered free at some time. I am happy that this is the time. Again, thank you for supporting us.

Beth Sufian answers questions from the readers regarding cessation of work, medication co-pays and the FMLA in "Ask The Attorney". The spirit of caregivers is **Isabel Stenzel Byrnes'** topic in "Spirit Medicine". As usual, "Information from the Internet", compiled by **Laura Tillman,** is filled with helpful items for our perusal.

The Focus topic of this issue is: Gastrointestinal Issues. **Jessica Newport** writes of her experiences with GI problems after lung transplant. **Ana Stenzel** tells of her bouts of gastrointestinal cancer. The theme continues with **Kathy Russell** writing of her twisted intestines. The topic is rounded out in "Wellness" where **Julie Desch** examines the GI system from the mouth to the other end.

Learn about Spin class from **Jennifer Hale** in "Coughing With A Smile". **Nicole Matthews** is featured in "In The Spotlight". The "Poetry Corner" has **Jeanie Hanley** reflecting on her relationship with her sister, who also had CF. See **Ana Stenzel's** review of a new book, "My Foreign Cities", on page 18.

A helpful resource for genetic information is highlighted on page 7. An exhibit from "Through the Looking Glass" is on page 15. Find information about the BEF's "You Cannot Fail" program on page 21. Check out "CF Online" on page 28. As always, there are photos "From Our Family Album" on pages 16 and 17.

We would love to receive stories from you. Look at "Looking Ahead", on page 3, to see if there are any future Focus topics that interest you. We can assist you if you are unsure of your writing skills. We're happy to help.

Stay healthy and happy,
Kathy
Managing Editor

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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**

Spring (current) 2013: Gastrointestinal Issues.

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.) Do you live with pain or have you had to live with pain? If so, how do (or did) you handle it? Do you try to eradicate it or do you learn to live with it? What tips can you share with the readers?

Winter (February) 2014: Memory Problems. (Submissions due December 15, 2013.)

ASK THE ATTORNEY

Questions From the Readers, Answers From the Attorney

By Beth Sufian, JD

The past three months have brought many questions about determining when to stop work, co-pay assistance and the FMLA. Nothing in this column is meant to be legal advice about your specific situation. If you have additional questions please contact the CF Legal Information Hotline at 1-800-622-0385. The Hotline provides free and confidential legal information to people with CF, their families and their CF Care Center teams. The Hotline is proudly sponsored through a grant from the CF Foundation. Callers will speak to an attorney employed by the Hotline. The Hotline is the only such service that provides legal information from attorneys who focus their practice on the rights of people with CF. The Hotline also can be reached by e-mail at: CFlegal@cff.org.

> *"If people wait too long to stop work they may find it difficult or impossible to improve their health once they do stop work."*

BETH SUFIAN

Question 1: When Should I Stop Work?

Many adults with CF work in a variety of jobs. Some people with CF can work for many years while others find that, at some point, working has a negative effect on their health and they must stop work to spend more time taking care of their health.

There is no magic pulmonary function number that indicates a person should stop work. People should determine if their work schedules prevent them from performing necessary medical treatments, exercising and getting enough rest. If there is no way to take care of oneself while working full time, alternatives to full time work should be explored. If people wait too long to stop work they may find it difficult or impossible to improve their health once they do stop work. Honest consultation with the treating CF physician is important. Many CF physicians do not realize how much time it takes to perform daily medical treatments and may underestimate the role fatigue plays in the ability of a person with CF to work. It is important to convey to one's CF physician the effects on healthcare that a person has in making time for work.

Social Security benefits are the only federal government benefits offered to a person who can no longer work due to disability. Often people with CF think there are many programs to provide income support if a person is unable to work. The Food Stamp program and the HUD housing program may help with assistance with food and housing for people with CF who live in low income households. However, the only federal programs offered to provide income support due to inability to work because of a medical condition are the Social Security Disability Insurance (SSDI) and Supplemental Security Income (SSI) programs.

Congress has been holding hearings recently about the increase in the number of Americans receiving Social Security Disability and SSI benefits. Many in Congress think that too many people receive benefits and see cutting the benefit rolls as a way to help balance the country's budget. The Social Security Administration (SSA) has stepped up efforts to review the cases of individuals receiving SSI benefits and SSDI benefits.

Many people with CF have never had a SSA Continuing Disability Review (CDR). Typically, CDRs happen every 3-5 years for a person with CF. However, recently, many people with CF have been reviewed sooner, especially if the person is working part time. There is no way to avoid a CDR once notification is received that a CDR for a particular person has been ordered by the SSA. There are ways

to make sure that, at the end of a CDR, SSA will agree that benefits should continue.

People with CF should make sure that they attend their CF Center Clinic visits regularly and that they have PFTs and sputum cultures performed at each visit. Medical records are a very important piece of evidence in a CDR. Non-compliance with prescribed medical treatments is cause for termination of Social Security benefits. People with CF should make sure they adhere to their prescribed treatments.

People with CF who choose to work part time while receiving benefits should make sure that their monthly work activity does not violate the Social Security work rules. First, for a person who receives SSDI, work activity cannot exceed $1040 a month from part time work activity. In addition, the person should not be working full time even if the monthly income is below $1040. A good rule of thumb is to not work more than 4 hours per day, 2-3 times a week, while making less than $1040 a month. Working 4 days a week for 8 hours a day could result in termination of benefits, even if the monthly income generated from such work activity is below $1040. This is because so much work activity is close to working full time and the SSA could find that a person who can work that much could work a little more and, thus, is able to work full time. A person is not allowed to work full time and receive SSDI benefits.

While a person on SSDI can have nine trial work period months (typically it is only nine months in a person's lifetime) where he can make over $1040 a month, it is not advisable. Those who are working making more than $1040 as a trial work period month are finding that the SSA will conduct a Continuing Disability review and, more likely than not, will find the person is able to work full time and will stop benefits. If a person with CF is not ready to go back to work full time he should carefully consider the idea of using his trial work period months. He should understand that even though a person is allowed nine trial work months, the SSA can decide at any time that a person is not eligible for benefits and terminate benefits.

Second, a person who receives SSI benefits will have different work rules and income limits. SSI work rules are very specific and will depend on the amount of the benefit check a person is receiving. Some people with CF who receive SSI do not receive a full benefit check and, therefore, the amount the person can make from work activity will be less than a person who receives a full SSI check. There are specific work programs that allow people to work and continue to receive either an SSI benefit and Medicaid or just Medicaid. In most states, without SSI, an adult with CF CANNOT receive Medicaid even if the person with CF has no other way to obtain health insurance or will not be able to obtain medications. If the adult does not have SSI he will lose his Medicaid. Once a person goes over the allowable amount all benefits stop, including Medicaid.

This column has contained this information in the past but, now, it is even more important to make sure you understand the work rules for the Social Security benefit program from which you are receiving benefits. Contact the CF Legal Information Hotline at 1-800-622-0385 to discuss SSI or SSDI work rules and work programs.

Question 2: I cannot afford my co-pays for medications. Is there anyone who can help?

The CF Patient Assistance Foundation (CFPAF) provides co-pay assistance for certain medications. The CFPAF can be reached at 1-888-315-4154. The CFPAF is a subsidiary of the CF Foundation. However, the CFPAF does not provide assistance for all medications taken by a person with CF. A person must apply for assistance from a variety of assistance programs for medications that are not covered by the CFPAF. A person may have to fill out a lot of forms for each drug for which the person needs co-pay assistance. Most CF Centers have staff members who can assist you with the application process.

Question 3: Can I take FMLA leave and then take vacation time and sick leave?

An employer can run FMLA leave concurrent with vacation time, sick leave and short term disability or long term disability benefits. For example, if Jane is sick and needs to be hospitalized for three weeks, she can request FMLA leave if: she has worked for her employer for one year prior to requesting the leave, she has worked 1250 hours in the past year, and her employer has 50 or more employees in all offices within a 75 mile radius. However, her employer can require her to use her two weeks of vacation time and one week of sick time concurrently with FMLA time. After she is out for three weeks, she would then have nine weeks FMLA time remaining. ▲

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.

SPIRIT MEDICINE

The Spirit of Caregiving

By Isabel Stenzel Byrnes

Like most people with CF, I have been the recipient of caregiving all of my life. In fact, I owe my life to my caregivers! My parents have helped me, nurses gave me my meds, respiratory therapists pounded my chest… the list goes on. As we adults with CF get older, we are not immune from taking on caregiving roles to people important in our lives: dependent children, an aging parent, a sick sibling or spouse (with or without CF). Even if you aren't in a caregiving role today, at some point you may be. In this article, I'd like to explore the art of caregiving from the perspective of someone living with CF.

What is a caregiver? For purposes of this article, a caregiver is a family member or friend who helps someone in need, who is not able to fully function independently due to illness or age. To be a caregiver is a decision. For most of us it is instinctual, born of genuine love for another person, whose wellbeing matters to us. For some people, caregiving is an obligation, because there's no alternative helper, and that moves them to action. Reciprocity drives some people; a grown child may return the care he received to an elderly parent. As an on-again and off-again caregiver for my very independent sister, Ana, who is undergoing chemotherapy (see her article, CF and Cancer – The Gift that Keeps on Giving on page 14,) I know helping my twin sister would be mutual: she'd be there for me if I were in her shoes.

Having CF complicates the caregiving instinct. I'm reminded of a silly teenager at CF camp who used to walk around yelling, "All the attention on me! All the attention on me!" There was something true about her statement. CF usually centralizes a focus on us. Our inner child might welcome the one-way street of being cared for. As someone who was raised as a care-recipient, I admit there is a part of me that is afraid to take on too much of a caregiving role. Being a caregiver takes time and it is easy to lose focus on our own CF needs. Besides practical things, worrying about someone, thinking of their appointments schedule, or their longevity, can take the mental focus away from our own needs. It's hard to juggle both. My most honest thought is: "I can't take on all the responsibility; I have CF – I can't overdo it or I'll get sick." I was raised with a heavy conscience that certain decisions might rock the fragile balance between health and sickness, energy expenditure and saving.

> *"Caregiving reminds us that we are all inter-connected; that what we do for others matters to ourselves."*

ISABEL STENZEL BYRNES

I'd like to go deeper with this issue. Caregiving may reveal less pleasant parts of our humanity. I bet it would be challenging to take care of someone who does not want to care for herself, or who feels entitled to be cared for. Caregiving won't always be reciprocated, leading to the inevitable question, "Who will care for me?" During my caregiving experience, I've encountered a range of negative feelings. I feel guilty for not doing more. I feel guilty for not wanting to be the caregiver, as I have enough on my plate. I feel guilty someone else is suffering more than me. I notice resentment when illness cancels plans, or when someone doesn't listen to what I think would be helpful for her. I even feel envy when friends and family focus solely on Ana's health. And caregiving is easiest when there is a timeframe: there is a beginning, middle and end; prolonged neediness can be draining—even if it's not intentional.

There are many positive aspects to caregiving – including the spiritual and interpersonal benefits of caring. A fun-

damental human requirement as social animals is to get along in adverse circumstances; to still love and care unconditionally for someone despite stress, mood changes, even physical repugnance. Nearly all people, including myself, hold an innate capacity to be caring, loving, and supportive to another person in an intimate way. We have evolved this way. The spiritual source of caregiving should not be fear-based, like there will be some divine punishment or karmic judgment if we don't take care of our loved ones. A real consequence of caregiving is a feeling of satisfaction, purpose and worth, which raises self-concept. Personally, I believe caregiving is life-saving. I have seen many CF adults gain a greater will to live when they have children, telling me, "I must survive because this person needs me." Caregiving reminds us that we are all inter-connected; that what we do for others matters to ourselves.

Caregiving is also enlightening, and helps me be a better person. To fully focus on another person, to be present and attentive, is healing because I can get outside of my own experience for once. I can practice patience, non-judgment and the gift of companionship. It reminds me I am one member of a team and that I have a role in delegating, asking for support, and reducing unnecessary tasks. Being a caregiver helps me prioritize what's really important – my loved ones. Caregiving also reminds me that boundaries are important. I have a tendency to nag my sister so she'll do what I think is right. Though it's well-intentioned, it is annoying for both of us. I am my own person. I must respect my sister's space and support her independence. If she wants to do something by herself, I bless that. Just because I try to help her out here and there doesn't give me dominion over her choices, even if I don't support them. I also learned that what she is going through is not necessarily what I will go through. The person who needs my help is having her own unique experience that I will never fully understand, even as a caregiver and witness. So no matter what, the experience is lonely.

To reverse roles and focus on another person is not always easy. Yet I believe people with CF are at an advantage. We make great advocates for our medically-fragile loved ones, because we are incredibly medically savvy and know how to communicate in the medical setting. More importantly, we know what it feels like to receive care, so we have a special sensitivity to providing it. And maybe our situation reveals a basic truth. Osho, in his book *The Virtues of Selfishness: Love, Freedom, Aloneness: The Koran of Relationships*, says that to be altruistic, we must be selfish. He writes, "Only a very deeply selfish person can be unselfish. A self-centered person is always seeking his happiness. And this is the beauty of it, that the more you seek your happiness the more you will help others to be happy. Because that is the only way to be happy in the world." Though we *have to* be selfish and focus on our own health, we can be better caregivers by doing so.

If you are a person with CF who is also a caregiver, I imagine there are inner resources you use to keep going. I hope you have found balance in your caregiving role that is fulfilling. You are welcome to share them with me, or write to *CF Roundtable* at: cfroundtable@usacfa.org. ▲

Isabel Stenzel Byrnes is 41 and has CF. She lives in Redwood City, CA with her husband, Andrew. You may contact her at isabear27@hotmail.com.

Genetic Mutation Information Resource

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

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

SPEEDING PAST 50...

As The Intestines Turn

By Kathy Russell

It looks as though spring is on the way. Where I live, in NW Oregon, we have had a mild winter and are already seeing signs of spring. The forsythia and violets are blooming and so are some of the rhododendrons. The daffodils are coming right along, too. I am so happy to have made it through another winter – unscathed. My husband made it to March before he caught some rotten upper respiratory bug. So far, I have not caught that bug and I am keeping my distance from him, in hopes of avoiding it.

The Focus topic of this issue is "Gastrointestinal Issues". I am too familiar with this topic. Some of my earliest memories are of having trouble with my gastrointestinal system. I think I always have had reflux; I just didn't know what it was until I got older. I remember how eating certain foods would cause my throat to feel as if it were on fire. Others caused my stomach to burn or caused me to have a lot of burping and/or gas. (I don't even want to go there!)

I learned which foods to avoid, pretty quickly. I knew that I couldn't eat chicken fat. If it just touched my esophagus, I would suffer with terrible pain and burning. Even seeing the yellow fat that floats on the top of a can of chicken noodle soup, makes me feel ill. I can't stand the feel of it in my mouth, either.

While I'm mentioning fats, I should explain that I cannot tolerate the feel of most fats in my mouth. The tropical oils are really unpleasant to me. If I eat something of the type of "Cool Whip", I can't get rid of the feeling that my mouth is coated in paraffin. It is a truly waxy feel. Coconut oil is the only tropical oil that doesn't seem to cause me such discomfort.

I am careful with all fats. I can tolerate small amounts of cow's milk, cream, cheeses or butter. I use milk that is no fatter than 2% and use cream or butter in very small amounts. I use sunflower seed oil or olive oil for cooking. Neither of those causes me pain. I don't use canola oil because it leaves a "fishy" taste in my mouth. I use pan sprays for much of my cooking.

There are other foods that cause me discomfort. I avoid canned tomato sauce. For some reason, it causes burning in my esophagus. Other types of tomato products don't bother me, so I use them instead of sauce. I must be careful about combining tomato and oil. Fresh tomatoes with oil are fine, as are canned tomatoes and oil. Tomato paste or canned sauces mixed with oil create a volatile mixture. I have likened it to pouring gasoline down my throat – and then lighting it! No thanks. I'll pass.

KATHY RUSSELL

As long as I can remember, I have had a problem with food not moving through my system in a normal fashion. Hours after I had eaten something, if I were to 'burp', I would get a mouthful of the food that I had eaten hours before. Of course, this was accompanied by a lot of acid. It caused a lot of burning in my throat. I had a lot of scarring and tenderness in my esophagus. I was diagnosed with GERD (gastro-esophageal reflux disease).

A few years ago, after having G-I surgery, I began taking Nexium®. This has been a real help to me. I no longer have to put up with GERD. Also, food seems to move through my system in a more normal way now.

Getting food to move through me has been wonderful. I no longer have that constant feeling of being over full in my stomach and needing to have a bowel movement. I should note that I have added a daily dose of polyethylene glycol – a generic form of Miralax – to my daily regimen. Between that and the Nexium, my innards are working quite well.

I don't miss the terrible pain that I used to feel in my abdomen. It could wake me from a sound sleep, and it did – frequently. It would feel as though someone had punched me in the gut. If I was awake and standing up, it would double me over. There were times that it felt as if my stomach or part of my intestines had just flipped over. Usually, a short time later, I would feel a 'reverse flip' and the pain would ease. A few times, the reverse flip didn't happen. I would be in terrible pain for a long time.

When I was awakened with really intense pain, I found that the only comfortable position was on my elbows and knees with a hot water

bottle held to my abdomen. I would let my head hang down and try to relax. I found that the contoured cushions from an old couch made a great support for my aching innards. I would just get on that old couch, bent over the cushion and try to rest. After a few hours, the pain would ease and I could go back to sleep. Since my last abdominal surgery, I haven't had a recurrence of that pain.

Since I've mentioned surgery, I guess I should explain it a little. I have had a couple of abdominal surgeries because of CF. The first one was necessitated by an intestinal abscess. It was a very large abscess, about the size of a grapefruit. It had pushed my bladder out of its way and was causing me intense pain. I ate a little dinner the day before I went to the hospital and didn't eat again for three weeks… and there still was stool in my intestines! (These CF innards are weird.) I had an exploratory laparoscopy with removal of my gall bladder and half of my large intestine, which also is known as the colon. (I guess that means that I now have a semi-colon!) The surgeon said that my small intestine had a few "kinks". He straightened those out. He warned that I would most likely experience that kind of kinking again - and I have. One good thing about the surgery - he said that there was no evidence of any cancer in my abdomen. Since my mother had colon cancer, this was a great thing to hear.

My next surgery was because of an obstruction and a kinked small intestine. Foolishly, I had eaten dry-roasted peanuts without enough added lubrication. My mouth always has been rather dry. I can't eat peanut butter without jam, jelly or honey to lubricate it. Evidently, I can't eat peanuts in the same way. Anyway, I developed a lot of pain and I was starting to vomit. (I rarely vomit, so that was notable.)

I was hospitalized. My surgeon and I decided to wait and see if my kinked small intestine would unkink on its own and let the obstruction move. It didn't. After waiting for a week, he went in and moved the obstruction manually and unkinked the intestine. He had taken all of my intestines out of the cavity to unkink them as well as cleaning them of adhesions. He said that my entire abdomen was full of adhesions and that the removal of the adhesions took longer than the rest of the surgery. Once again, there was no evidence of cancer.

My earliest surgery that was related to CF guts was when I was 23. I had severe hemorrhoids with a rectal prolapse. I did not know that I had either problem, since that was the way it always had been. No one had ever asked me about either problem and, until I had significant rectal bleeding, I didn't know that there was anything wrong. Once I healed up, I felt much better and was much more aware of such problems.

I have had long periods of taking supplemental enzymes and long periods of not taking them. Currently, I am not taking them. I seem to be getting along just fine without them. I do not have a problem with holding weight. In fact, I have the exact opposite problem.

I come from a family that has generations of morbid obesity. I believe that works in my favor. Although I am not morbidly obese, I am overweight. When I get ill and have a fever, I can lose ten pounds in a few days. If I were at my ideal weight, this could prove problematic. Since I always carry extra weight, I can afford to lose ten or twenty pounds without significant problems. Thanks be to the genes of my ancestors.

I wish that I had some words of wisdom to share with you about how to live with CF guts. Sadly, I don't. All I know is that our innards are quirky and can cause us great pain and problems. I hope that you can get a good system working for you and that you will have fewer problems than some of us have had.

Eat well, exercise wisely and think positively. ▲

Kathy is 68 and has CF. She is the Managing Editor of CF Roundtable. She and her husband, Paul, 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

Ed Fleischman
Plainview, NY
71 on December 24, 2012

Arthur Herron
Sacramento, CA
32 on September 13, 2012

Wedding

Kathy & Paul Russell
Gresham, OR
48 years on March 27, 2013

Transplant

William H. Coon, Jr., 53
Spring, TX
Bilateral lungs
4 years on January 28, 2013

NEW BEGINNINGS

Grandparent

Paul Feld, 55
Florissant, MO
Became a grandfather
On March 26, 2013
When his daughter, Sarah
Gave birth to a son,
Christopher James

Information from the Internet...

Compiled by Laura Tillman

SINUS

Endoscopic Sinus Surgery in Adults with Cystic Fibrosis: Effect on Lung Function, Intravenous Antibiotic Use, and Hospitalization Endoscopic Sinus Surgery in Adults With CF. Oswaldo A. Henriquez, MD; Lindy L. Wolfenden, MD; Arlene Stecenko, MD; John M. DelGaudio, MD; Sarah K. Wise, MD. Arch Otolaryngol Head Neck Surg. 2012;138(12):1167-1170.

The study aims to ascertain the effect of endoscopic sinus surgery (ESS) on lung function, intravenous (IV) antibiotic use, and hospitalization in adults with cystic fibrosis. This preliminary study of ESS in adult cystic fibrosis patients indicates significant reduction in the number of inpatient hospital days in the postoperative period. However, there is no evidence that ESS improved lung function or the need for IV antibiotics.
http://tinyurl.com/ck4wxr4

FYI

Long-term multicentre randomised controlled study of high frequency chest wall oscillation versus positive expiratory pressure mask in cystic fibrosis. Maggie Patricia McIlwaine, Nancy Alarie, George F Davidson, Larry C Lands, Felix Ratjen, Ruth Milner, Blythe Owen, Jennifer L Agnew Thorax. Published Online First: 13 February 2013

This study was done to determine the long–term efficacy of high frequency chest wall oscillation (HFCWO) compared with positive expiratory pressure (PEP) mask therapy in the treatment of cystic fibrosis as measured by the number of pulmonary exacerbations (PEs). The results of this study favor PEP and do not support the use of HFCWO as the primary form of airway clearance in patients with CF.
http://tinyurl.com/b5hpzwf

Left ventricular and aortic dysfunction in cystic fibrosis mice. Zachary M. Sellers, Attila Kovacs, Carla J. Weinheimer, Philip M. Best. Journal of Cystic Fibrosis. Published online 26 December 2012.

In a mouse model of CF, CFTR mutation leads to left ventricular remodeling with alteration of cardiac and aortic functions in the absence of lung disease. As CF patients live longer, more active lives, their risk for cardiovascular disease should be considered.
http://tinyurl.com/c7ubhk2

Baby bottle steam sterilizers disinfect home nebulizers inoculated with bacterial respiratory pathogens. Dana Towle, Deborah A. Callan, Patricia A. Farrel, Marie E. Egan, Thomas S. Murray. Journal of Cystic Fibrosis. Published online 26 December 2012.

Continued on page 19

Mailbox

My sister has cystic fibrosis. She is 46 and is a reader of *CF Roundtable*. I find it very informative! I am a Respiratory Therapist at a local hospital.

Stephanie Altmeyer
Richland, IN

Thank you for all your hard work and dedication!

Anonymous

Keep up the good work!

James Yankaskas, MD
Chapel Hill, NC

Thank you for all your work. I really appreciate *CF Roundtable*.

Darleen Boynton
Ann Arbor, MI

The board must receive congratulations from each of us fortunate enough to have been blessed by your growing *CF Roundtable*.

Joan P. Tilney
Yarmouth, ME

FELD *continued from page 1*

child, and don't even bother asking for a second in the future.

So, on March 4, 1983, we drove to the agency to pick up our daughter, Sarah. It was her first birthday, but she became our present for life. Sarah has been through a lot with me. She has witnessed countless physical therapies, hospital stays and, ultimately, my transplant over eight years ago.

She had also endured my divorce eight years after her adoption. While both my first wife and I remarried relatively fast, our custody agreement meant that, until Sarah graduated from high school, she would live one week with me and the next with her mom. That created all sorts of issues; but I am certain it most heavily weighed on Sarah, who had an on/off button to switch every week as she moved between us. It's certainly not a life I'd wish on anyone.

Regardless of these challenges, both my first wife and I have been happily married to new people for many, many years. My marriage will reach 22 years in June. During college, Sarah mostly lived on campus or a short distance away in an apartment. College for her was close to Kansas City, and I lived in St. Louis, so we did not get to see each other as often as I would have liked. Sarah had lived in the Kansas City area from 2003-2012. She met her future husband, Christopher, at a Royals game. They had a short courtship and were married in October, 2010. A little over a year ago they moved to the St. Louis area. Last September, she announced she was pregnant - due March 30, 2013.

I often speak at classes at Washington University in St. Louis with my transplant surgeon. The topic is lung transplantation. My surgeon talks about the history of the procedure and goes into some detail on ethics, survival, organ donation, etc. I then conclude the lecture with my personal story. I talk a lot about cystic fibrosis, the roadblocks in my life, and how blessed I have been to have lived this long with CF.

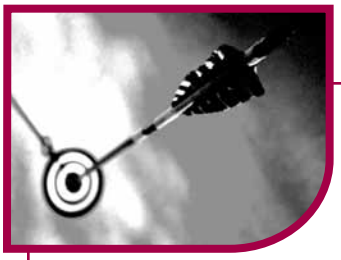
I am fairly certain that at 55, I have outlived at least 98% of patients with CF. A lot of my friends and family have called me a miracle man, and I have almost died a couple times. I am not comfortable with that adjective, but I will admit to being a man of strong faith and one who complies strictly with what my physicians tell me to do.

So, on March 26, 2013 (just last week) Sarah gave birth to her first child, a boy named Christopher James. Both mom and baby are doing great. Both grandma and grandpa are doing great. Both step-grandma and step-grandpa are doing great. I have found it amazing how a child can strengthen the love in a blended family, and everyone is willing to pitch in and help when and where they can.

I can comfortably say I have a "CF family of friends" that numbers around 100 people whom I know fairly well. In that group, there might be one other CFer who is a grandma. That puts me in some very rare company. It is certainly a blessing, if not a miracle, and something I never expected. I'm going to ride this wave as long as possible!

For all you CFers out there who are consumed by statistics about the disease, there always are stories like mine that can provide hope for you and your family and friends. My physician was right in saying, "I have no idea how long Paul will live, only God knows." This grandpa is grateful for his blessings, of which I've had many. I hope many of you can repeat this story in your own fashion in future years and, if you have made it to 'grandma' or 'grandpa', *CF Roundtable* would love to hear from you with your story.▲

Paul is 55 and has CF. He is a Director of USACFA. His contact information is on page 2.



FOCUS TOPIC

GASTROINTESTINAL ISSUES

Gastro-WHAT? A Journey after Transplant

By Jessica Newport

As with most CFers, my pancreas is enzyme-insufficient, so I have taken enzymes since my diagnosis when I was 21 months old. Despite having CF, my digestive system was always pretty sympathetic to me; no feeding tubes, no blockages, no problem! Just the usual stomach/intestinal pains if I didn't take enough enzymes.

However, my lungs were not so cooperative. Due to my decreasing lung function, I had a bilateral lung transplant on April 18, 2010 at the age of 23. I had a feeding tube put in after the surgery because the doctors had deemed it necessary to avoid eating by mouth until I could have a Nissen fundoplication; a standard procedure at the hospital. The "fundo" as I call it, though it is no fun, keeps food from aspirating into the new lungs. Sometimes when a person coughs or chokes, some food can bypass the epiglottis and get into the lungs. Usually, this causes a massive choking episode; thus the phrase "went down the wrong pipe". I understood why I needed to have the surgery, but it couldn't be scheduled until a month after my transplant.

While waiting on my fundo, I experienced some real issues with tube-feeding through a gastric port. The major one was that, about an hour after starting the feed, I would inevitably throw up. No anti-nausea medicine worked. It made the entire feed a nightmare, so I would hurry up and try to fall asleep before the nausea kicked in. After the fundo surgery, I was still using my tube because the doctors wanted to keep me off of solid foods for a few weeks.

JESSICA NEWPORT

I was so excited to start eating again! However, nausea and a new symptom, pain, occurred after nearly every meal. I attributed the symptoms to not being quite ready for that much solid food. But the nausea and pain continued. For one of my weekly appointments I was told not to eat for 24 hours. I had a chest x-ray taken, and my doctor told me he could see the slice of pizza I had eaten over 24 hours ago. A stomach-emptying test (radioactive eggs test) was done to see how fast my stomach emptied over time. The results: after one hour, my stomach had only emptied 11%.

> "The "fundo" as I call it, though it is no fun, keeps food from aspirating into the new lungs."

Now, my family and I were concerned and confused. None of this had happened before my transplant. My parents, being worried, searched for hours on line trying to figure out why this was occurring. My mom finally came across the answer…gastroparesis. It translates to paralysis of the stomach. During certain surgeries, an important nerve called the vagus nerve can be accidently affected or cut. More research on our part told us the whole story.

Gastroparesis can often happen to lung transplant patients during surgery, especially CF patients, because the vagus nerve is located along the spine where our lungs have to sometimes be "scraped" out due to all the mucus and years of infections. Unfortunately, I was one of those "lucky" patients. It takes the stomach an average of 4 to 5 hours to empty after a meal. My stomach emptying results weren't even close. Diabetes can also induce slower emptying of the stomach and gastroparesis has been found quite frequently in CF patients who had not had a transplant. Thanks, mucus!

There are certain pills that can help speed up the stomach emptying process. Erythromycin and Metoclopamide (Reglan) can certainly help, but also carry side effects. As a result of my continuing stomach-emptying problem, I had a common procedure for gastroparesis called a pyloroplasty done. The pyloroplasty

involved the widening of the entrance to the small intestine from the stomach, so that food can pass more easily into the colon. However, another radioactive eggs test post-surgery confirmed little improvement.

For me, all I do now is try to drink as much liquid as possible to help move things along. Diet is key, if the medications do not work. I cut out soda, which has helped, and haven't had a vomiting incident in over a year. I still get the occasional cramping from a big meal, but I don't actively think about gastroparesis anymore and it's been almost three years.

I'm not trying to scare anyone who is thinking about transplant, but merely informing all about what can happen. We were never told about the likelihood of gastroparesis beforehand, so I would hate to see someone else surprised by it. Things turned out fine for me, as it will for you! ▲

Jessica is 26 and has CF. She is three years post-transplant. She lives in Durham, NC. You may contact her at: nu6586@tampabay.rr.com.

In Memory

Gregory Baxter, 42
San Diego, CA
Died on January 29, 2013

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

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

CF and Cancer - The Gift that Keeps on Giving

By Ana Stenzel

I've always joked about all the things that CF really stands for- **C**oughing **F**it, **C**ontrol **F**reak, **C**onstantly **F**arting, **C**omplains **F**requently, **C**lose **F**riends. But never in my wildest dreams did I ever imagine that CF would, for me, stand for Cancer Fighter.

In 2011, at the age of 39, I was diagnosed with small bowel adenocarcinoma. I had learned at a CFRI conference years ago that people who carry the CF gene have an increased risk of gastrointestinal cancers. After all, my mother and about three other CF carrier parents that I know had battled colon cancer. Even a decade after my lung transplant, the CF gene, with all its sneaky mischief, proved to be the gift that keeps on giving – this time by giving me the Big C.

So when my GI symptoms drastically changed in February of 2009, I suspected cancer. Almost overnight, my digestive system went on strike. No matter how many enzymes, probiotics, anti-acids or laxatives I took, my symptoms were not resolved. I changed enzymes four times, to no avail. I would go to the bathroom up to nine times-a-day then be in the ER with a bowel obstruction the next day– it made no sense. For two years, I was in chronic pain which disrupted sleep, exercise, intimacy and my social calendar. Pain also affected my posture, my gait, my travel schedule and my enjoyment of my hobbies such as hiking and swimming. My CF and lung transplant physicians referred me to a gastrointestinal doctor who proceeded to run every test she could think of for me - endoscopy, colonoscopy, CT scan, gastric emptying test, stool anaylsis, upper GI study and many more humiliating and unpleasant tests. I had more gastrogaffin enemas and CF cocktail enemas than I could count. Nothing showed any suspicion of cancer. My weight bounced up and down like a yo-yo and

ANA STENZEL

I became terrified of certain foods that triggered obstructions such as red meat, spinach and granola. My belly was so distended that I was asked if I was pregnant. My twin sister, Isabel, became frustrated that our travels together often involved numerous urgent quests for a bathroom, and her patience was tested.

In July 2011, with my weight and energy at an all time low, I told my mother, "I'm going to die of CF gut disease" because I didn't see any solution to this downward spiral. My low hematocrit (indicating anemia) and my GI symptoms were all attributed to my transplant meds or being "old" with CF. That made no sense to me since my identical twin, who also had CF, would take her enzymes and Miralax and be just fine. It all seemed so hopeless and unfair.

Then, after a particularly agonizing hospital stay for another bowel blockage where I was without any food, TPN or IV nutrition for five days, I demanded to be discharged from Stanford Hospital to attend the world premiere of our documentary film about CF and lung transplantation, *The Power of Two*. Our film was showing at DocuWeeks, a premiere film showcase in Los Angeles. Within hours of arriving, having eaten two small meals of soft foods, I was doubled over in pain again and unable to move. I went to USC Hospital Emergency room and had another CT scan which showed nothing. After three days of excruciating pain and a belly that made horrific rhythmic noises like a screeching washing machine, I consented to exploratory surgery.

The surgeon found a baseball size mass in my small intestine - he described it like a meatball in a pile of spaghetti. It was cancer. I knew it! My mother, whose adenocarcinoma of the colon was diagnosed 15 years earlier, also had a low hematocrit. Her tumor was on the colon side of the ileum; my tumor was on the small intestine side of the ileum. I was discharged nine days later from USC and 20 pounds lighter. I have never experienced such severe pain as I had after my bowel surgery. It made the two lung transplants that I had in 2000 and 2007 feel painless compared to this.

Back at Stanford, I expressed my disappointment that my cancer was

Continued on page 26

PHOTO BY STEPHEN BOYER

Pill

You wanna come inside my mouth?
You dirty mind,
my tongue is dirtier. these pills
are bitter. Gelatin capsules really
aren't that scary...
How's the weather out there?
How are your kids? Your life?
Your dog? What color are
my teeth ?

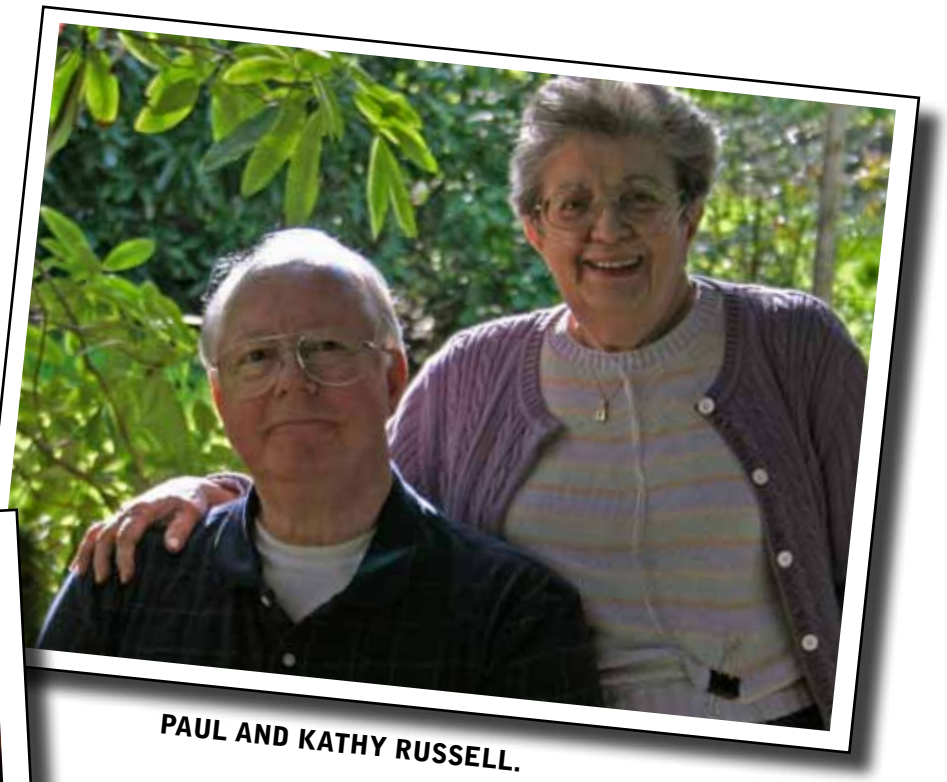
What is the
color of humility?
man, I'm bored. let's Keep
this train moving. move on
to a more interesting denial.
If I close my teeth and
grin, do these pills disappear?

Then quit pretending.

-Todd G., 2002

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

FROM OUR FAMILY PHOTO ALBUM...



PAUL AND KATHY RUSSELL.

MICHAEL KOWAL AND NICOLE MATTHEWS AT THE "CYSTIC FIBROSIS RISING STARS" DINNER. NICOLE WAS AWARDED A FOLLOW A RAISING STAR AWARD.

JESSICA NEWPORT

ISA STENZEL BYRNES, ANDREA EISENMAN AND ANA STENZEL POSE IN TIMES SQUARE, NEW YORK CITY, DURING A SNOWFALL.

BOTTOM CENTER ARE JOHN AND JEANIE HANLEY, PARENTS OF, FROM LEFT, MARIA, KEVIN AND JESSICA.

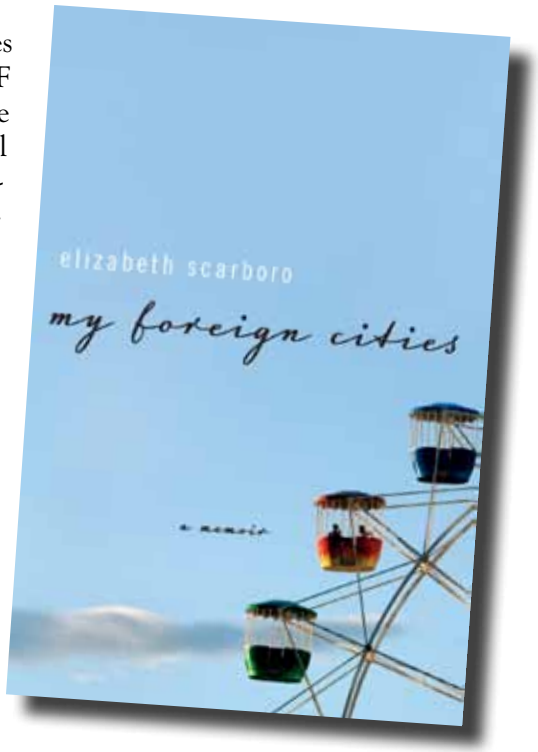
BOOK REVIEW

My Foreign Cities

A Much Needed Read for the CF Community

By Elizabeth Scarboro
Reviewed by Anabel Stenzel

Elizabeth Scarboro's memoir, My *Foreign Cities*, is a much needed book for the CF community. Sharing her experiences as a young woman who falls in love, while in her late teens, with a man who has CF, and their 10 year journey together as life partners moved me and provided insights into the experience of the CF spouse. In all the literature I've read on CF, rarely have I come across a story so powerful and insightful in providing the spouse's viewpoint on the CF experience. The book is beautifully written, humorous at many points and so real at others, and gives the reader an in-depth personal account of the joys and frustrations of loving a spouse who happens to have CF. Like some CF books, I did not leave the experience feeling down, critical or frustrated but, rather, infused with love and appreciation for those who are able to love people with CF unconditionally – "for better or for worse, in sickness and in health." Elizabeth's writing style brings you deeply into her experience and emotions and allows the reader to get to know her husband with CF, Stephen, in an intimate way. Through story telling based on memory, she touches upon important issues facing CF adults such as: the fine balance between pursuing career/educational goals and health, the risks of addiction from prescription pain medications, the desire to delay hospitalizations because of knowing all too well the song-and-dance red tape that awaits once admitted, and that final surrender to medical care when illness takes over and there is no choice.



This is a story of young love and devotion of two best friends-turned-lovers surviving the challenges of young adulthood together despite the horrendous nature of cystic fibrosis. As more adults with CF are fortunate enough to find supportive life partners, this book is essential to allow us to recognize and appreciate their story, their experience and the joys and tears of loving someone with CF until the end and beyond.

As a person with CF who is married, I found myself in Stephen and could only imagine the shared frustrations and joys that my own spouse experiences. I felt a brotherhood and kindred spirit in Stephen, for he, like many of us with CF, share some common characteristics brought about from being raised with CF. I, like Stephen, can be obstinate, highly determined, possess a keen self awareness of my physical well-being, and am all too aware of my limited time on earth. Like so many of us with CF, Stephen grabbed onto life and cherished every day, even if it meant putting off that visit to the ER, or minimizing those CF hassles like hemoptysis and lung collapses, despite the concern of those around him. More than once I found myself with Stephen, and

Be sure to check us out on:

Twitter: http://twitter.com/#!/cfroundtable
Facebook: http://www.facebook.com/CFRoundtable
Blogspot: http://www.cfroundtable.com/blog-2/

TILLMAN *continued from page 10*

probably would have chosen the same course of action as he, as we balance our health, educational and job pursuits, and family needs.

Only through reading *My Foreign Cities* – did I truly grasp how frustrating, yet strangely comical, it must be to witness the choices made by people with CF from our loved ones' perspectives. In my experience in relationships, I've always thought how much a CF relationship can be a bit one sided or lopsided, with so many needs and plans arranged to fit the CF spouse's health and medical schedule. Elizabeth eloquently shares that reality in an honest yet accepting way but also points to the valuable life lessons that she received despite some sacrifice. How fortunate we are that our spouses love us not only despite CF but that they also so lovingly (and amazingly) accept so much of the lifestyle and personality traits that go along with it. Yet, through Elizabeth's writing, it is clear that she, like many spouses, is profoundly impacted by loving someone with CF and that Stephen, like many people with CF, had so much to offer those who loved him.

I truly recommend this book for all partners, spouses or spouses-to-be of any illness community, so that we can have greater awareness of the personal struggles and joys of our partners. You may laugh, you may cry, you may reflect on your own experience. Most likely, you will come away with appreciation that love is not chosen – it just happens – and when it does, it lasts forever.

For more information, contact Elizabeth Scarboro via Facebook at: https://www.facebook.com/myforeigncities?fref=ts. ▲

Anabel Stenzel is 41 and has CF. She is a genetic counselor and an author of: "The Power of Two: A Twin Triumph over Cystic Fibrosis". She lives in Redwood City, California, with her husband and dog.

Contaminated nebulizers are a potential source of bacterial infection but no single method is universally accepted for disinfection. All steam sterilizers used in this study were effective at disinfecting all home nebulizers. Viable bacteria were not recovered from any inoculated site after steam treatment, under any conditions tested. Steam treatment is an effective disinfection method. Additional studies are needed to confirm whether these results are applicable to the clinical setting.
http://tinyurl.com/bo8uv7k

Peripheral neuropathy in cystic fibrosis: A prevalence study. Biswaroop Chakrabarty, S.K. Kabra, Sheffali Gulati, G.S. Toteja, Rakesh Lodha, Madhulika Kabra, R.M. Pandey, Achal Srivastava. Journal of Cystic Fibrosis. Published online: 07 February 2013

Patients with cystic fibrosis have electrophysiological evidence of peripheral neuropathy (predominantly axonal, sensory and polyneuropathy). There is significant association of higher chronological age with occurrence of peripheral neuropathy.
http://tinyurl.com/b8ozxjn

Dynamic vascular changes following intravenous antibiotics in patients with cystic fibrosis. James H. Hull, Rachel Garrod, Timothy B. Ho, Ronald K. Knight, John R. Cockcroft, Dennis J. Shale, Charlotte E. Bolton. Journal of Cystic Fibrosis. Volume 12, Issue 2 , Pages 125-129, March 2013

Adults with cystic fibrosis (CF) have altered large artery haemodynamics which is associated with a persisting systemic inflammatory state. The abnormal central haemodynamics evident in adults with CF is modulated with a short intervention of intravenous antibiotics.
http://tinyurl.com/azgzpks

Adherence to Airway Clearance Therapies by Adult Cystic Fibrosis Patients. Theodore Dassios, Anna Katelari, Stavros Doudounakis, Gabriel Dimitriou. Respiratory Care, Published online: 27 February 2013.

The study was to determine rates of self–reported adherence to airway clearance therapy (ACT) by patients treated in an adult cystic fibrosis program, to identify patient characteristics associated with poor adherence, to typify adherence according to ACT technique, and to indicate reasons for poor adherence. Study outcomes showed a high rate of ACT adherence in adult CF subjects. Lower level of education was the most important factor in poor adherence to ACT. Self–reported adherence and treatment recommendations were in best agreement with positive expiratory pressure and flutter device techniques.
http://tinyurl.com/bozw2u6

Aerobic exercise and respiratory muscle strength in patients with cystic fibrosis. Theodore Dassios, Anna Katelari, Stavros Doudounakis, Gabriel Dimitriou. Respiratory Care. Published online: 27 February 2013.

The beneficial role of exercise in maintaining health in patients with cystic fibrosis (CF) is well described. The authors' objective was to compare respiratory muscle function measures in CF patients who regularly exercise with those CF patients who do not. The results suggest that CF patients who undertake regular aerobic exercise maintain higher indices of respiratory muscle strength and lower pressure time index of the respiratory muscles values, while increased upper arm muscle area values in exercising patients highlight the importance of muscular competence in respiratory muscle function in this population.
http://tinyurl.com/bozw2u6

PRESS RELEASES

Celtaxsys Initiates Phase 1 Study of

Continued on page 28

COUGHING WITH A SMILE...

Come Spin With Me,

Come Spin With Me, Come Spin Away With Me

By Jennifer Hale

Hello *CF Roundtable* Readers! Hope you are enjoying the spring season that is upon us now. I wanted to share with you, in this issue, my recent experience at Spin class. For those of you who do not know what Spin is, I will explain. It is fancy stationary bicycles that are designed with resistance knobs, handle bars that can be moved up or down, and peddles that hold your feet in so you can stand while riding. They are really cool and they give you a great work out!

With my oxygen (O_2) tank in tow, I set up my bike the other day to start Spin class. Setting up is a process of adjusting the seat, the handle bars and resistance. Along with wiping the bike down with sanitary wipes and then setting up my O_2 tank so it does not fall, Gatorade goes in the drink spot and a towel is kept handy. I have found that just hanging my O_2 tank bag on the handle bars works best for me. I do have a back pack bag I can put the tank in and I do use that when I am biking outdoors, but I do not like to use it on the Spin bike because the less weight I have to carry on my back, the easier it is to ride. Since the back pack is on your back (hence the name) and the O_2 tanks are not the lightest, whenever I can I use my over the shoulder bag. I hang it on the handlebars if that is convenient or sometimes, if I am lucky, it actually fits in the cup holder on the various exercise machines.

Anyhow… so I am all set up on the bike waiting for class to start. I have my O_2 fitted around my nose, though not turned on yet because I need every bit of O_2 in the tank to do the class. I am still self conscious sitting there with my O_2 and when I am out in the main gym with it on, too. But I have only gotten positive feedback from people who ask me about it. One piece of feedback I have gotten I will share with you in this story.

The instructor comes in and tells us we will be climbing today. That means lots of resistance on the bike and lots of visualization of climbing a mountain. Climb every mountain, la, la. Okay I digress! LOL! What I love about Spin is the visualization and the way you can go at your own speed. The visualization is wonderful because I can motivate myself by visualizing that I am climbing that mountain and the goal is to get to the top! I'm King of the world! LOL!

I find I visualize a lot in my ways of dealing with CF. I tend to think of happy places when I am going through tough times or tough procedures. I feel the power of visualization is very strong and very healing. I also like that you can Spin at your own speed. This is great for people who are out of shape, have compromised lung function or anything that would limit them in working out. I can peddle the bike at my own speed and put my own resistance and no one knows I am not doing it at the level of the teacher or anyone else. I am doing it at my speed and at my level but still getting a great workout. It really goes with my philosophy of dealing with CF. Some days are better than others and some periods of time are better than others. But I just keep adjusting my sails depending on my circumstances and I do the same at Spin class.

> *"What I love about Spin is the visualization and the way you can go at your own speed."*

JENNIFER HALE

Sometimes my resistance is more, sometimes I peddle faster and sometimes I stand up but, sometimes, I do not do any of that and just peddle in my saddle (sitting on the seat.) All that is okay! I am still out there, I am still doing it and I am still getting a benefit. I feel like in Spin it is not just about how hard you are riding or how

fast you are riding, it is about being there and doing it. Like the Nike saying, "Just Do It." So when I go I just do it to my level and enjoy participating in the class. I have to say, though, that I think I am getting old because I really thought the music last time was quite loud! LOL.

Well, the class was almost over the other day and the instructor was walking up and down the aisles chanting her motivational words of wisdom when she stopped by me and pointed to her nose and asked me, in the middle of class, on her microphone, why I have the oxygen. I thought I would be mortified but, really, it was my chance to say why and then people can move on and not look at me and wonder. Although, I am sure no one was even paying attention. But when something is different about us, I think we all feel self-conscious to some degree, because we all just want to be accepted for who we are. Right?

So I tell her I have cystic fibrosis and only a 40% lung capacity. She says in the microphone how I am her hero and how strong I am - yadda, yadda, yadda. Then the guy next to me starts nodding his head in agreement. I thought I was going to fall off my bike. My feet were fastened into the shoe straps so that would have been impossible. Ha, ha! But, really, I was relieved and now it was not such an elephant in the room.

Again, her reaction was fantastic and it is the third time I have gotten a response like that in the gym. People say how strong I am, but I do not feel that what I am doing is strong. I feel it just is what I have to do to stay healthy.

I tell myself that if one day I have to use the big tanks and roll my O_2 tank into the gym, I will do so. But it is not easy. It is not easy feeling like you are different. It is not easy to feel sad when you are working out because your lung capacity is not what it once was. It is not easy to huff and puff even though you have O_2 cranking in your nose. The workout is still tough on my lungs and I still labor to breathe. It is not easy being green, LOL.

But it is what it is. That is one of my favorite sayings. It seems to really sum up certain situations. I cannot control that I have to use oxygen, but I can control getting my butt off the couch and continuing to work out and be active even though I have had to make adjustments.

That is what life is all about: making adjustments. When you are dealing with an illness that limits you, it is about making the most of what you can do and not about what you cannot do. Are some days better than others? Oh yea! There are some days that I just cannot work out and that is okay. Like the quote in one of my favorite movies, *Gone with the Wind*, "Tomorrow is another day!" So get out there. Start small and do what you can, not what you can't. Even if it's a two-minute walk to the mailbox, build on that and set small goals. Dealing with CF you will constantly have to start over with your goals because getting an exacerbation really throws a wrench into trying to work out - but that is okay. As Tom Hanks says in the movie Castaway, "Tomorrow the sun will rise and who knows what the tide will bring."

Until next time my friends! ▲

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

YOU CANNOT FAIL

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

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

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

WELLNESS

Just Put My Ashes On The Back Of The Toilet:

That's Where I Spend Most Of My Life

By Julie Desch, MD

These were the words spoken by my brother, Tom, when we discussed his wishes around my parents' kitchen table a few years before he died of CF at age 58. Tom had a wry sense of humor, but as many of us know, there was quite a bit of truth hidden within his joke. True, the man could not eat enough Velveeta cheese, and perhaps this contributed some to his dilemma, but I completely understood his point. CF is not just a lung disease, as it is commonly defined. It wreaks havoc on the gastrointestinal system as well, but I probably don't need to tell this to you.

> *"Even if you were lucky enough to be born without meconium ileus, there still is a chance to experience its adult cousin, DIOS."*

The lungs get all the press, but the effects of CF on the GI tract can be a major pain in the butt, pun intended. At 52, I've dealt with a few common issues myself, but one of the benefits of writing this article is realizing just how many I haven't (yet?) had the pleasure of experiencing. In the resultant spirit of gratitude, I present "THE CF GUT", starting from the mouth and working south.

I have been somewhat perplexed, but mostly amused, by the occasional trick my salivary glands play on me. As they are epithelial structures, with glands and ducts and all of those things that are messed up in CF, it is not surprising that they sporadically act up. There I will sit, minding my own business, when my tongue will make a swipe of the inside of my mouth, and will come up with a strange white material that, when I touch it, has the consistency of rubber cement. This is of no consequence, of course (other than the entertainment value), but it does point out what is happening in pancreatic ducts, biliary tree, and sadly, the lungs.

Moving down the esophagus, GERD anyone? This stands for **g**astro-**e**sophageal **r**eflux **d**isease, and is the result of stomach contents (food and liquid) backing up into the esophagus. This stuff is very acidic, and the esophagus is not made to tolerate such insult, so the result is heartburn, burping, nausea, and other unpleasantries. This can even damage the esophagus, leading to atypical change in the cells near the junction with the stomach. This is termed "Barrett's" esophagus, and must be monitored closely as these cells are considered precancerous.

JULIE DESCH, MD

Descending further down the alimentary canal, the small intestine provides a few potential challenges to those who live with CF. First; the mega-big problem that presents at birth is meconium ileus. Meconium is the name of the first stool that a newborn has—I'm not sure why it gets a name and subsequent stools don't. Even in normal babies, this stuff is thick and sticky. In CF, it is thicker and stickier, to the point where it can't budge and creates a roadblock in the ileum (the last part of the small intestine). The result is pain, abdominal distension, vomiting, and a very sad and uncomfortable baby. If you had this, you might have a scar from the surgery to remove the blockage…although sometimes it can be successfully treated with enema flushing.

Another possible issue arising in the small bowel is termed SIBO—small intestinal bacterial overgrowth. Normally, the first half of the small bowel has very small numbers of bacteria and the epithelial surface of the entire small bowel is not "colonized," meaning the bacteria might be there in small groups within the lumen, but

they have not set up shop. In SIBO, there are too many bacteria and/or they are of abnormal type within the small bowel. This doesn't happen just in CF, but risk factors include the use of proton pump inhibitors (treatment for GERD), pancreatic insufficiency (present in 80% of people with CF), and previous small bowel resection (occasionally the treatment now, more commonly in the past, for meconium ileus). So we are more at risk than the average Joe. Here's the problem: the symptoms of SIBO are gas, bloating, diarrhea and/or constipation, and pain. Sound familiar? Exactly. These are very non-specific symptoms that are common to all of us with absorption issues.

So…let's move down to the colon, shall we? Even if you were lucky enough to be born without meconium ileus, there still is a chance to experience its adult cousin, DIOS (distal intestinal obstruction syndrome). Known fondly as "a blockage," DIOS is not at all uncommon. The same issues that lead to meconium ileus (weird mucus and abnormally dehydrated epithelial surface) combine with other factors including malabsorption of fat and systemic dehydration to plug up the best of CF bowels. I shall heretofore name the result of this process, "rubber cement poop." The obstruction usually occurs at the junction of the small intestine with the large intestine, AKA, the cecum. This is a bit of a cruel trick, because the cecum is right where the appendix lives, providing a diagnostic dilemma for doctors not familiar with cystic fibrosis bowel problems. Appendicitis = surgery, whereas DIOS usually flushes well with a rotor rooter technique, and surgery is rarely required.

A less common problem is intussusception of the bowel. Intussusception is one of my favorite bowel-related words…right up there with borborygmi (go ahead, Google it). Imagine your colon is a telescope: intussusception is simply when part of your colon becomes enveloped in the adjacent part—like when a telescope folds up. This needs to be treated immediately, as it can lead to necrosis of the bowel and significant infection. Often times, the procedure used to diagnose an intussusception corrects it—a barium enema.

Finally, we come to the end of the colon, the rectum and anus. Rectal prolapse is no fun, according to my brother. Rectal "prolapse" occurs when the rectum, which normally abides inside the body, decides to wander out and see what's up, or in this case, down. This is actually not that rare in kids with CF, and is thought to arise due to coughing spasms (increasing intra-abdominal pressure) combined with large amounts of rubber cement poop. In us old folks, you can add years of sitting on the pot straining as an additional risk factor.

Even if you have escaped rectal prolapse, you still may be very familiar with its distant cousin, hemorrhoids. In fact, if you live long enough with CF, this is almost a guarantee. Hemorrhoids are just very swollen blood vessels (veins) that line the rectum and anus—the tail end of the GI tract. These can hurt, they can itch, and they often bleed. If they are located higher up, they may be asymptomatic (other than the blood), until they too, decide they need to come out to play—and prolapse. Sigh. A very painful complication is when one becomes thrombosed. When this happens, you feel like you are sitting on a nail. Ouch. Go see your doctor if this happens…relief is just a scalpel away.

But wait, there's more. There are two very large glands that dump their contents into the small bowel, and are therefore very important players in the gastrointestinal system. You know these glands…the pancreas and the liver. These, too, are messed up in CF. In fact, the pancreas is so messed up that cystic fibrosis was actually named after the look of CF patients' pancreases. They were cystic (think Swiss cheese) and fibrotic (scarred). Rubber cement-like secretions block the pancreatic ducts, and these secretions, being enzymes, basically digest away pancreatic tissue. The result – the need for enzyme supplements to digest food. Another result can be potential ongoing pancreatitis (chronic) or sudden bouts (acute) of pancreatitis leading to pain, nausea, vomiting, etc.

Loss of pancreatic exocrine (enzyme producing) tissue is often accompanied by loss of the endocrine (insulin producing) cells as well. This absolute loss, plus abnormal insulin secretion in the first place (a CFTR issue), plus possible abnormal insulin sensitivity in the cells of the body can lead to a very special form of diabetes – CF-related diabetes, or CFRD. The prevalence of CFRD increases with age, so that by the time we reach middle age, one in every two Senior Cystics is reaching for insulin shots on a daily basis.

Rubber cement-like bile secretions in the biliary tree cause a similar problem in the CF liver. After the liver cells make bile, it is dumped into a complex system of small ducts that eventually lead to the small intestine. There, it emulsifies fat and assists in its digestion and absorption. But rubber cement bile gets stuck, causing inflammation, which causes scarring and (usually) slow destruction of liver tissue. Sound familiar? This leads to the hallmark CF liver finding (exciting for us pathologists) of focal biliary cirrhosis—this is a fancy name for bile duct centered scarring of the liver which is patchy in nature, associated

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IN THE SPOTLIGHT

With Nicole Matthews

By Andrea Eisenman with Jeanie Hanley

Nicole is our featured guest and has willingly shared her amazing young life with us. She has so much energy and we hope her story so far energizes you! In addition to having CF, rheumatoid arthritis and other health issues, she has graduated from college, recently moved into a new place with her boyfriend, works full-time, teaches ice skating, volunteers as an EMT and most recently published a new book, *The Abnormal Nicole*, which in our opinion, is a beautiful story of love and endurance. Please welcome our latest star – Spotlight please!

SISTERS KATIE AND NICOLE MATTHEWS.

Age: 24

Occupation: Reputation Manager for West Herr Toyota and Scion in Orchard Park, New York

What is a Reputation Manager?

I work full time and sometimes more! My face is the first face that people see when they enter our dealership. I greet them, offer a cup of coffee or tea and get the customers settled in wherever they need to go – sales manager, consultant, service, parts, wherever they need. I handle a lot of correspondence and am in charge of getting reviews for the website. I also handle the payroll and am in charge of the Simonize (protective coating for cars) appointments, along with ordering coffee supplies and brochures for the store.

Work-related Challenges and Tissues

I'm blessed with this job. I was hired before my uncle/godfather, who is the owner, knew I was applying, so nepotism was not a factor. It turns out that the people here are very understanding. I'm an open book about my life so they have always been aware of the CF and my other health problems like rheumatoid arthritis, scoliosis and gluten allergy. They give me a break whenever I need it. I keep a box of tissues at my desk, along with a ton of hand sanitizer. I am able to read whenever I need a break, or just a breather. Because of their support, I rarely miss work. Also, friends at work participate in Great Strides every year.

The Areas Affected Most by CF and Life Expectancy

CF affects mostly my digestion, but I also use the Vest®, Pulmozyme® and albuterol and have had several sinus surgeries. When I was diagnosed as a baby, at 22 months, it was the stomach bloating and endless amount of diaper changing that led my parents to understand that something was wrong. An elevated sweat chloride confirmed CF. My mother, who is an RN, checked the medical books, all of which stated that the life expectancy was 18 years. Although my parents were very sad at first, they decided that they wanted me to live a very full life. After my little sister (Katie) was born, we took many trips, lugging around medications and treatments everywhere we went.

Enzymes, High School and Please No More!

Like others with pancreatic problems, I must take enzymes, Creon 20. The only time I was hospitalized was for DIOS when I was 16. In high school, I found out that I would lose weight if I didn't take the enzymes. I wanted guys to like me! Also, I had to go to the school nurse for the enzymes and missed out on lunch with my peers a lot, so I had another incentive not to take them. It became so bad that the doctors planned on inserting a feeding tube if I didn't straighten out. I realized then that I couldn't help others without helping myself first. I wanted to be successful in life and so I started taking the enzymes.

About two years ago, I was diagnosed with a gluten allergy. This almost pushed me over the edge. I had blisters

on my face, chest, lower stomach, armpits, elbows, scalp, and buttocks. (It is called dermatitis herpetiformis.) These blisters itched horribly, and truly wore me out. I had to cut out many foods and lost weight initially, but now it's under better control.

Religious or spiritual?

I believe in God and that I've been given these problems because I'm strong enough to handle them. I live to the fullest every day and if given a chance to live life over again, so far I wouldn't change a thing… except for the gluten allergy! I want to be able to eat everything! I believe that as a result of the CF and my spiritual beliefs, that I have a different outlook on life. Little things make me very happy – for instance, when my new business cards arrived and whenever I see a butterfly.

After my grandmother died of ovarian cancer at age 59, I noticed butterflies everywhere and they became a symbol of her. Soon after I got a butterfly tattoo to remind me that she is always with me. Often when I'm at my lowest point or during important turning points in my life, I'll see a butterfly. One day I walked out for a breather after a bad day at work and a butterfly had landed on my car window, giving me hope and faith that better days were to come.

What was it like growing up with CF when your sibling didn't have it?

When we were growing up, we were jealous of each other. I was envious that she was healthy and didn't have to visit the nurse at school or go to doctor's visits so often. She saw me as being catered to and getting all the attention. Even through this jealousy we always were close and she still is my best friend. Although she is 3 ½ years younger, she is taller and more muscular. I refer to her as my bodyguard and a true blessing.

BOCES and Blessings in Disguise

During elementary school the teachers were not very understanding at all with my CF. We had many conflicts with medications and sick days. School was frustrating at first, but as I went on in my school career, it became much easier and people were much more understanding.

During high school I received a BOCES (Board of Cooperative Educational Services) in Animal Care. I had planned to work with animals and for three years after graduating, I worked at a kennel. While working there, I needed sinus surgery and they refused to give me time off. I had to quit eventually because my health suffered. Of course this turned out to be a blessing in disguise, because I found the ideal job at Toyota after I graduated from college.

Boyfriends, Dating and Teddy Bears

I met Michael through an online dating website. I had been in a relationship for five years before that but couldn't take the pessimistic outlook of my now ex-boyfriend. Michael is just the opposite! I initially tried hiding the CF and told the first guy on the website about it and he said outright that he didn't want to date a sick person.

Well, I wanted to date an optimistic person. When Michael came along I informed him first off. He was a positive thinker and understanding. He has been phenomenal. He sits up with me during my midnight coughing fits and understands when I have an upset stomach. We've been living together since November 2011. He reminds me to do my treatments and helps with the little things. For my December birthday, he gave me two Teddy-Bear's wearing t-shirt's saying, "Please cure CF because I love my girlfriend" and "Hug me. I'm not contagious", along with two pillows saying "CF Sucks" and "When a cure is found we will be free" with a purple butterfly in the background (that symbol of my grandmother all over again).

Living Spaces and Mind Over Matter

Michael and I live in an 1821 farmhouse apartment in New York. Since moving in, my PFTs have improved and I am healthier by taking a "mind over matter" approach. I walk a mile in the morning with Michael, appreciate my surroundings, listening to birds chirping and just loving the wooded area. My two cats, Manasseh and Sophie, live with us along with our hedgehog, Hank. I wasn't allowed to have a dog, but the bulldog is still up at my parents' house in Boston, New York. The CF Center is close by in Buffalo and my family lives within the same town.

In *CF Roundtable,* you've submitted writings and poems. What inspired you to write?

One of my passions is writing and poetry. I've been writing all my life. It is my escape and also allows me to look at things differently, have a perspective. In fact, I've published a book titled, "The Abnormal Nicole" which has been available since April 2012. The main message is this – no matter what life throws at you, you can overcome anything. That theme is also embodied in my favorite saying: Life is like a rose, there are thorns, but it's still beautiful.

Hobbies – EMT, Ice Skating and ATVs!

My favorites are my work as a volunteer EMT, ice skating instructor and riding ATVs. As a volunteer EMT, I get paged and am able to help people of all ages. I'm careful to let others deal with patients who have respi-

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ratory illness and to not dive into fires, so that I can maintain my health. As a volunteer ice skating instructor, I teach kids and adults with all sorts of disabilities. My sister, father, and I had become volunteers teaching the "Gliding Stars" an adaptive ice skating program. We also volunteer with SABAH (Skating Athletes Bold At Heart). I started as a beginner and eventually advanced to a volunteer position.

Do your boyfriend and family members have as much energy as you?

My boyfriend says, "You tire me out!" My energy probably is my dad's influence. He can't say no and neither can I. He skates, participates in CF fundraisers, is also a firefighter, works full-time for the highway as a deputy and much more. My mom also does not say no to anything; we are a family that is always willing to help out, and go – go - go!

Who is your inspiration?

My friend Christine who passed away from CF. Knowing her and her family has inspired me to live even a fuller life, and to take care of myself. I would also have to say my family inspires me every day, even in the littlest of ways.

Where do you see yourself in five years?

I plan to be married, living in my own house, having kids and still be working full time.

What do you hope people take away from your life, your book?

I hope people realize that you may have these hurdles, and you've jumped over a lot of them, but live life to the fullest. Each breath is a blessing. We are meant to help other people, in some way. Don't worry – you can do it. And know that with all the volunteers and researchers working for us, we will see a cure in the future. ▲

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, please contact either Andrea or Jeanie.

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missed. I now had stage 4 cancer and was referred to oncology, a depressing, crowded cattle-car clinic with a whole different feel than my beloved CF and transplant clinics. I did six months of chemotherapy, of two drugs - one is appropriately named 5-FU, and the other is oxaloplatin which caused me to lose sensation in my feet. The chemo was not bad - it was like doing home IVs for two days every two weeks. I did not lose my hair or appetite. I was never nauseated. I joked that I had the kidney transplant version of chemo.

The cancer clinic and infusion center were always crowded with older, frail people, who looked and acted miserable. Some were just unlucky like me; others had invited their cancers by lifestyle choices. Most were unfamiliar with how to be sick. Unlike CF clinic or transplant clinic, there was no friendliness or desire to share in each other's journey. One nurse coordinator of the GI Clinic was extremely cold and unempathic and speaking to her was like speaking to someone who hated her job. This experience really reinforced to me how fortunate we are in the CF world, where we have a disease that is rare enough so that we have a bit more attention and affection from our healthcare providers and CF peers. I was so used to coming to CF /transplant clinic and having a familiar face greet me with a cheerful, "Hi!! How ya doing?!" So it was a true shock to be in the cancer clinic where I was greeted by a stranger with a straight face saying nothing but "Name and Date of birth?"

Cancer is extremely common and CF is relatively rare. Perhaps I was spoiled by the loving, long term relationships for this relatively small patient population at CF/transplant clinic…or maybe CF patients are really special and our CF providers recognize that and treat us accordingly. I'm not sure. Even when I post online on CF websites or Facebook pages, I get a response IMMEDIATELY since CF patients care about each other. I posted many times on cancer sites trying to find another person with small bowel cancer to talk to and never got a single response. The CF community is special.

I completed chemotherapy in six months while taking time off work. I stayed home and drank Peptamen (nutritional supplement with medium chain fatty acids for easier fat digestion) and lots of milkshakes and ate Costco croissants and guacamole. Finally, my weight was returning. By April 2012, I was almost back to my baseline weight and I returned to work. I became manager of Team Northern California for the 2012 Transplant Games of America and worked on getting back in shape.

However, my belly pain returned within four months of ending chemo. A follow up CT scan, done at my request, revealed more swollen lymph nodes suggestive of metastases. Nonetheless, I was determined to go to the Transplant Games and compete. I ran track and swam despite my numb feet. Amazingly, I won medals in every

event I entered. I was so happy to be alive and good nutrition was a huge part of that.

On August 31, 2012, exactly one year since my last surgery, I had bowel surgery again. The cancer had metastasized, turning my right ovary into a 9 cm mass, and leaving tumors all over my belly like "sand sprinkled" as my doctor described. The ovarian mass did not show up on CT scan! They removed another foot of small bowel and my right ovary. Surgery recovery was easier, probably because I was in such great physical shape from the Games. One month later, I started chemo again, this time with more 5-FU and a new drug, Irinotecan, which is nicknamed, "I run to the can" because of the side effects.

I was back on disability again, and spending time researching second opinions and alternative treatments. I am a forward-thinker so I was preparing Plan B and C in case this chemo didn't work. I read about alternative therapies in Germany, where there are many other cancer treatments that are not approved of in the USA. Several sources I read stated that there are ways to cure cancer, but those are kept from American patients, because it is a multi-billion dollar industry and profit motive keeps patients from being truly cured. How dreadful! Also, I read that conventional cancer therapies include a "cut, burn and poison" approach which often has more risk of killing the patient than the cancer itself. I was determined to consider alternative means of regaining my health by being educated, proactive and thinking outside the box.

After three months of chemo, a CT scan showed more possible metastases. My spirit was crushed. Despite colon cancer being the #2 cancer killer in America, there are only three drugs for it and options were running out. I am disillusioned and not surprised that so many people die from cancer. Unlike CF, you can't tell when the drugs are working. You have to wait 3-6 months to see (by a questionable scan) if chemo is doing anything. If you are lucky, the treatment won't kill you. Unlike CF, cancer is a passive disease, and you can't know or feel exactly what is happening within. For the first time, I feared dying from this since being on immunosuppressive medication for my transplant is a direct contradiction to fighting cancer. But I quickly dismissed these thoughts and pressed on. My oncologist said I would probably not qualify for any clinical trials for experimental drugs because of my CF and lung transplant. Instead of accepting his opinion, I researched Clinicaltrials.gov and found a trial at MD Anderson Cancer Center in Houston, Texas that I am pursuing.

I contacted several physicians in Germany to inquire about alternative treatments, but almost all are immune boosting to fight cancer, and that would likely put me right into rejection. I may come to a point where I have to choose if I wish to die of cancer or rejection. Which would you chose? I decided that if I can die with healthy lungs, then I would die happy.

To cope with cancer, I'm doing acupuncture for pain, eating anti-cancer foods, doing relaxation and imagery, pray, and I watch a lot of comedy. I also got a puppy which gives me new joy and purpose. I surround myself with love and enjoy food while I can. I returned to work, despite chemo, for my mental and financial health. My gut is still very dysfunctional, but I've found a way to still be happy and love life. I dream of the day of having a normal bowel movement - it's been years.

I know reading this article can be scary – after all, isn't CF enough? Our society poses such a huge stigma on the word "cancer" and it can instigate huge fear.

For me, fighting CF has given me the skills to fight cancer. I never passively accept the doctor's word (after all, it is most often just an opinion), and I believe no one will decide when I will die except me and God, Darwin lives in the medical setting. I am assertive and proactive with my care, and most of all, I know that life can still be active and happy despite illness. I try to live in the moment, not dwelling on the future, though I am realistic. Undoubtedly the lessons learned from CF will make my cancer journey and the journey of others dealing with CF and cancer, easier. I encourage you all to talk to your CF doctors about the increased risk of GI cancers, ask for appropriate screening, especially if you have a sudden change of bowel habits and a family history. This advice applies to CF carriers *and* CF patients. I recently learned that some CF centers are doing colonoscopies on all CFers over 40, using a special CF bowel prep protocol since conventional colonoscopy protocols don't always work for CF. Most importantly, it is important to recognize that not all gastrointestinal distress for us can be blamed on CF. That was the biggest mistake of my healthcare team. I hope that by reading this article, another CF family member will be spared from stage 4 metastatic cancer and years of pain.

Resources: There have been some reports suggesting that adults with CF carry a higher risk for carcinoma. A 1992 study reviewing 28,511 patients with CF found a nine-fold increase in risk for digestive tract malignancies (esophagus, stomach, small and large intestine, liver, and biliary tract) and a 30-fold increase in risk for pancreatic malignancies. This finding has been supported by several cases in the literature of pancreatic cancer in CF patients. In patients with young-onset pancreatic cancer (aged 42–50 years) there was an 8.4% (14 of 166) carrier rate of delta F508 compared with a 4.1% carrier rate in the control population.

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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.

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Anabel Stenzel is 41 and has CF. She is a genetic counselor and an author of: "The Power of Two: A Twin Triumph over Cystic Fibrosis". She lives in Redwood City, California, with her husband and dog.

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with abnormally regenerated liver tissue. More significant biliary cirrhosis is fortunately much less common, but can be a significant problem, usually in kids. When you hear of a CF brother or sister who needed a liver transplant, this is usually why.

Much more common liver problems include abnormally elevated enzymes, evidence of ongoing assault, and "fatty" liver for unknown reasons. Finally, many of us have gallbladder issues including gallstones and something called "microgallbladder" which is, as you might guess, a *teeny weeny* little gallbladder.

An article written by a pathologist would not be complete without a discussion of the C-word. Since we are all getting older, it is important to understand that we are at slightly increased risk for gastrointestinal cancers, including colon, small intestine, the gastroesophageal junction (remember this is where Barrett's esophagus develops), and the biliary tree. This risk is elevated following transplantation. So get your colonoscopies, people! ▲

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

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CTX-4430 Oral Treatment for Pulmonary Inflammation

CTX-4430 is a clinical-stage drug candidate designed to treat a broad range of chronic inflammatory disorders including cystic fibrosis. CTX-4430 works by inhibiting Leukotriene A4 Hydrolase (LTA4H), the major rate-limiting step in Leukotriene B4 (LTB4) production. Orally administered, CTX-4430 has been shown to reduce LTB4 production and to improve clinical signs and symptoms of the inflammatory disease state. CTX-4430 has the potential to become a novel treatment that combines successfully with new and current therapies in pulmonary and other inflammatory disorders.
http://tinyurl.com/a6nxg2e

Phase 3 Results Announced for Aeroquin for Cystic Fibrosis

Aptalis announced the results of a global Phase 3 study of Aeroquin (levofloxacin inhalation solution)

POETRY CORNER

Together

By Jeanie Hanley

Ten years ago you passed to another realm,
Leaving me at the helm,
Unprepared to deal with the loss,
Depression hit at a great cost.
Many life events thereafter,
Chipped away at the laughter
Taking a greater toll than they should,
Declining health, disability, menopause –
not good.

Then I saw you, felt you, in my dreams,
Walked by your fav gladiolas near a stream.
Heard the AC/DC play,
Remembered your songs,
And the words you used to say,
We will always be together
And this is true,
A part of you lives within me,
And in this world too.

Many years passed
Before realizing the pain,
The depth of its presence,
Permeating my brain.
Then the need to treat it,
Get back up on the horse,
Trying many therapies to beat it,
And am now returned with a force.

The blues have lifted
As has the sorrow,
The joy has returned to my heart,
At least for today and hopefully many tomorrows!
The love that you showered will always be left.
This has been constant even after your last breath.

Dear sister, rest in peace,
The memory of you is alive and will never cease.
Our shared CF was just one small piece.
Thank you for leg wrestling, adventures and
funny faces,
For the laughing for no reason and spiritual places,
We will always be together
Now and forever.

Jeanie Hanley is a 50-year-old physician who has CF. She is a Director of USACFA and is the Vice President. Her contact information is on page 2.

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conducted in cystic fibrosis patients with stable chronic lung infection with *Pseudomonas aeruginosa*. Aeroquin is a proprietary formulation of levofloxacin for aerosol delivery using an investigational eFlow Nebulizer System. The trial consisted of three 28-day on/off treatment cycles. Patients had been heavily pretreated with inhaled antibiotics prior to enrollment. The primary endpoint – non-inferiority of relative change from baseline in percent predicted Forced Expiratory Volume in 1 Second (FEV_1) after the first treatment cycle of 28 days – was met. This effect with Aeroquin on lung function was maintained over all three treatment cycles (ie, through Day 168).
http://tinyurl.com/atkwlf8

KaloBios Initiates Phase 2 Study with KB001-A Humaneered® Monoclonal Antibody in Cystic Fibrosis Patients

The study will investigate the safety and efficacy of intravenously administered KB001-A as a treatment for chronic *Pseudomonas aeruginosa* (Pa) infection. The primary endpoint of the study will be time to need for antibiotics to treat worsening of respiratory tract signs and symptoms. Secondary endpoints will include changes in inflammatory markers, respiratory

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symptoms, subject-reported outcomes, measurements of lung function, pharmacokinetics, safety, and tolerability. KB001-A is designed to neutralize *Pseudomonas aeruginosa* pathogenicity which reduces inflammation and allows the body›s natural immune system to kill and clear the bacteria.
http://tinyurl.com/ap5xly9

FDA Advisory Panel Rejects Cystic Fibrosis Treatment

A federal advisory panel has voted unanimously against the approval of dry powder mannitol (DPM) for the management of cystic fibrosis in patients aged 6 years and older to improve pulmonary function. The US Food and Drug Administration's (FDA's) Pulmonary-Allergy Drugs Advisory Committee's 14-0 vote against the approval sought by Pharmaxis for its novel inhalation product *Bronchitol* was based on a lack of data for significant efficacy combined with safety concerns, particularly about an increase in hemoptysis in children younger than 18 years.
http://tinyurl.com/akvg8ac

Study suggests cystic fibrosis affects nervous system

A new study by University of Iowa researchers suggests that the CF mutation also affects the nervous system and might directly cause some neural abnormalities experienced by people with CF. The researchers were able to show that the nervous system is directly affected by the genetic defect in CF. The UI team showed that the protein affected in CF is expressed and functions in a type of cell associated with nerves, called a Schwann cell. These cells produce myelin, a fatty substance that insulates nerve fibers and allows efficient transmission of nerve signals. The study shows that loss of CFTR protein directly alters Schwann cell function and leads to subtle structural abnormalities in the myelin surrounding the nerve fibers. These abnormalities, although significantly milder, resemble myelin defects seen in known human neuropathies.
http://tinyurl.com/b8yhmby

Grant Awarded to Study Novel Therapies for Muco-Obstructive Diseases Includes Studies on Cempra's Solithromycin (CEM-101) in a Cystic Fibrosis Anti-inflammatory Model.

Cempra, Inc. announced that the National Institute of Allergy and Infectious Diseases awarded a grant to Richard C. Boucher, M.D., Kenan Professor of Medicine and director of the Cystic Fibrosis and Pulmonary Research and Treatment Center at the University of North Carolina School of Medicine, to investigate the anti-inflammatory activity of solithromycin and its effect on mucin secretion in models for cystic fibrosis (CF). Solithromycin has demonstrated greater potency than azithromycin and other antibiotics against respiratory pathogens as well as stronger anti-inflammatory properties in vitro and in animal models.
http://tinyurl.com/b9vlhp9

Vertex Readies Late-Stage Cystic Fibrosis Drug Studies

The studies will evaluate the two different doses of an experimental medicine, VX-809, in combination with Kalydeco. Each study will enroll 500 cystic fibrosis patients randomized to either the VX-809/Kalydeco arms or a placebo for six months of treatment. The studies' primary endpoint will be the relative improvement in lung function of VX-809/Kalydeco compared to placebo. Last fall, Vertex demonstrated that a 600 mg dose of VX-809 and Kalydeco worked synergistically to improve lung function in cystic fibrosis patients with the F508del mutation compared to placebo. This same dose combination will be tested in the phase III study along with a higher 800 mg (actually, 400 mg given twice a day) dose of VX-809 plus Kalydeco. Along with the two phase III studies in adult patients, Vertex will also conduct a six-month study of the combination therapy in pediatric patients ages 6 to 11. If the data from the studies are positive, the drugs could be approved sooner than expected and for more patients. Vertex is conducting a phase II study of another experimental cystic fibrosis drug known as VX-661, which could be more potent than VX-809. Results are expected before the end of the second quarter.
http://tinyurl.com/a2rakv3

BACTERIA/INFECTIONS

Association between Staphylococcus aureus alone or combined with Pseudomonas aeruginosa and the clinical condition of patients with cystic fibrosis. Dominique Hubert, Hélène Réglier-Poupet, Isabelle Sermet-Gaudelus, Agnès Ferroni, Muriel Le Bourgeois, Pierre-Régis Burgel, Raphaël Serreau, Daniel Dusser, Claire Poyart, Joël Coste. Journal of Cystic Fibrosis. Published online: 07 January 2013

The prevalence of methicillin-resistant staphylococcus aureus (MRSA) in cystic fibrosis patients has increased and MRSA seems to be associated with a poorer prognosis. The aim of this study was to assess the prevalence and clinical consequences of MRSA and methicillin-susceptible staphylococcus aureus (MSSA), associated or not associated with Pseudomonas aeruginosa (PA). Clinical condition of CF patients with MSSA only or MRSA only appeared similar, whereas MRSA/PA patients had more severe respiratory function than MSSA/PA patients. In CF patients, MRSA might be more deleterious than MSSA only when associated with PA.
http://tinyurl.com/avlgr5p

Chronic *Stenotrophomonas maltophilia* infection and mortality or lung transplantation in cystic fibrosis

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TILLMAN *continued from page 30*

patients. Valerie Waters, Eshetu G. Atenafu, Annie Lu, Yvonne Yau, Elizabeth Tullis, Felix Ratjen. Journal of Cystic Fibrosis. Published online: 07 January 2013

Chronic *Stenotrophomonas maltophilia* infection is an independent risk factor for severe pulmonary exacerbations in cystic fibrosis patients. The goal of this study was to determine the effect of chronic S. maltophilia infection on mortality and the need for lung transplantation in a longitudinal study of children and adults with CF. The authors conclude that baseline chronic S. maltophilia infection is associated with an almost three-fold increased risk of death or lung transplant in CF patients. It is still unclear whether chronic S. *maltophilia* infection is simply a marker of severity of disease and ultimate mortality or whether it is causally related to disease progression.
http://tinyurl.com/a9w8ykn

TREATMENTS

A randomized controlled trial of inhaled l-Arginine in patients with cystic fibrosis. H. Grasemann, E. Tullis, F. Ratjen Journal of Cystic Fibrosis. Published online: 16 January 2013

Cystic fibrosis (CF) airways are nitric oxide (NO) deficient. The authors conclude that repeated inhalation of l-arginine in CF patients was safe and well tolerated. Inhaled l-arginine increased NO production without evidence for changes in airway inflammation.
http://tinyurl.com/az99f55 ▲

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

REMINDERS

- Please notify us immediately of any address changes. Returned mail wastes money and delays mailings.
- We would like to act as a referral source for active adult support groups. Please send us your group name, leader's name and phone number, number and age range of your members and geographical area covered, and we will add you to our referral list.
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- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq., 712 Main, Suite 2130, Houston, Texas 77005. Email: cflegal@cff.org**.
- You may subscribe at www.cfroundtable.com

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

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

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

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Helps defray out-of-pocket travel expenses for transplant recipients. Helps to set up trust funds. For more information, write to: AOTA, 21175 Tomball Parkway #194, Houston, TX 77070-1655

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