CF Roundtable

A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS AUTUMN 2011

Disaster Awareness in Cystic Fibrosis

Provided by Foundation Care (with special thanks to our current PharmD candidate, Aaron Arnold)

The recent weather disasters caused by large outbreaks of tornadoes bring to light the need to be prepared for such events. Those with chronic illnesses such as cystic fibrosis (CF) should also be prepared in case of an emergency or natural weather phenomenon. Many times these disasters come without warning and can eliminate many of the services that you depend on in order to manage your condition. This makes patients with CF very vulnerable during disasters.

The first thing to do is to identify what types of disasters are most likely to occur in your area. For those along the coast, it may be hurricanes or tsunamis while others may face floods, earthquakes and tornadoes. You can contact your local emergency management office or Red Cross for more information about disaster risks and plans for your area.

> "Many times these disasters come without warning and can eliminate many of the services that you depend on in order to manage your condition."

Next, make plans with your immediate family. Think about how you will reach each other and where you would meet if you were separated. Also, form an emergency support network. Your emergency support network may include teachers at school, a nanny, other family members, or friends. It is important to include out-of-town members in your emergency support network because those close by may be impacted by the disaster as well.

Your plans will require you to determine whether you are going to stay at home or leave the area. If you choose to stay, you need to consider:

- Items for daily living.
- Loss of utilities.
- How to get around.
- How to obtain additional supplies.
- How to get help.

If you choose to leave you need to consider:

- Everything above as well as the need to seek a new shelter.

If you must seek a new shelter, include in your plans shelters that are able to accommodate your condition. If possible design plans that minimize emergency shelters that may put you in contact with other infectious patients. Public shelters may not be

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INSIDE THIS ISSUE

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USACFA needs your tax-deductible donations to fund the printing, production and mailing costs of CF Roundtable. A yearly donation of $15 for individuals, $25 for non-U.S. addresses (U.S. funds only) and $25 for institutions is requested. However, CF Roundtable always is free of charge to those who are unable to donate. A fully completed subscription form is required to add your name to our mailing list. (If you have CF, please include your birth date.)

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

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EDITOR'S NOTES

The Focus topic of this issue is: "CF, It's Not Just For Children". This illustrates that people with CF are living much longer and healthier lives. Just in my lifetime, I have seen new therapies and advancements to treat CF that have allowed our median age of survival, once in the single digits, to rise to middle age. On our board of directors, some have reached retirement age.

Writing for the Focus topic, **Maria Fioccola** shares her experience of growing up with CF in a household that did not believe she had CF and how she overcame that by staying optimistic. Being diagnosed later in life is a theme from **Laura Mentch,** who writes about not knowing she had CF until she was 50 years old. Similarly, **Juliet Page** and **Kathleen Wilson** write about being diagnosed later in life, at ages 43 and 51, respectively. In "Speeding Past 50", **Kathy Russell** shares her tale of being diagnosed at 12 in 1956. She was continually told by disbelieving doctors, "You should be dead!" In spite of that, she is still going strong at age 67! In "Transplant Talk", **Andrea Eisenman** writes about her struggle with the median age of survival for CF.

Please welcome our new columnist, **Jennifer Hale,** who writes "Coughing With A Smile". She introduces herself and then discusses being an adult with CF and the responsibilities—staying on top of medical insurance, doctor appointments, medications and therapies. She feels that handling all of this is a job and she is the CEO - of herself. Our other new column is called, "In The Spotlight". It is written by **Jeanie Hanley** and me. Our first interviewees are **Anabel Stenzel** and **Isabel Stenzel Byrnes,** two vibrant examples of people with CF living life to its fullest. We are seeking others who have CF and are interested in sharing their stories with *CF Roundtable*. If you would like to be interviewed for this column, please contact Jeanie or me. Our contact information is to the left in the masthead.

In "Ask the Attorney", **Beth Sufian** discusses Social Security insurance notices of overpayment—what can be done, what ones' responsibilities are and the appeals process. In "Wellness", **Julie Desch** describes her perfect workout for any age, from rolling out her muscles, to exercises, to stretching. For "Information From The Internet", **Laura Tillman** culls the Internet for the latest news on CF–including new medications, advancements and treatments.

Recreation grants are available from the Cystic Fibrosis Lifestyle Foundation. (See page 27.) They provide recreation grants to help those who have CF live an active lifestyle and promote a healthier longer life through activities such as karate, golf, swimming, gymnastics, dance, horseback riding etc. Please contact them for more information.

Finally, I close with this reminder – please consult your doctors about staying current with the flu vaccine and ask about the pneumonia vaccine as well. Keep those lungs clear and healthy.

Stay well,
Andrea

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

Please share the milestones in your life with our readers. Your successes and achievements may serve as a source of motivation for others in need of an infusion of "positive mental attitude" in the pursuit of their goals. Send us a note specifying your "milestone." Include your name, age, address, and phone number. Mail to: ***CF Roundtable*, PO Box 1618, Gresham, OR 97030-0519. Or E-mail to: cfroundtable@usacfa.org**

ANNIVERSARIES

Birthday

Michelle Allen
Portland, OR
60 on September 14, 2011

Lucille Kinsel
Canton, IL
70 on July 10, 2011

Kathy Yoder
Portland, OR
49 on August 16, 2011

Wedding

Michelle & Gary Allen
Portland, OR
15 years on April 27, 2011

Andrea Eisenman & Steve Downey
New York, NY
3 years on September 13, 2011

Valerie & Rick Vandervort
Claremore, OK
20 years on June 14, 2011

LOOKING AHEAD

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

Autumn (current) 2011: CF, It's Not Just For Children.

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

Spring (May) 2012: Are You An Optimist or A Pessimist? (Submissions due February 15, 2012.) Do you see the glass as partly full or partly empty? Do you start each day with a smile or a moan? How does that way of thinking affect your health, your job, your family etc? Whether you are an optimist or pessimist, were you always that way or did you become that way? Does CF play a part in that?

Summer (August) 2012: Respiratory Therapy Activities. (Submissions due May 15, 2012.)

ASK THE ATTORNEY

Questions and Answers

By Beth Sufian, Esq.

The problems with our nation's economy are having an impact on people with CF and their families. The CF Legal Information Hotline has experienced a significant increase in calls from people with CF and their family members, especially in the past three months. Many of the individuals have lost their jobs and are in need of health insurance or government benefits. Some have found that their employers are less likely to allow them extra sick time if they need to be hospitalized, because the employers' staffs have been cut to a bare bones level. It is important for people with CF and their families to know which programs are available to help them access insurance coverage or, in certain cases, access medication without cost, until coverage can be found. Please contact the CF Legal Information Hotline for information on such programs.

In addition, as you have heard in the news, Congress is working hard to reduce government spending. The Social Security Administration (SSA) is under pressure to make sure that every recipient of Social Security benefits is, indeed, eligible for benefits. Social Security has stepped up its efforts to identify SSA benefit recipients who have worked and made over the SSA allowable amount. These individuals are not entitled to benefits once they have worked more than the allowable amount. Those individuals must pay back to SSA any benefit amount paid in a month when they were not eligible for benefits. This is called an SSA overpayment. This column will explain SSA overpayments and ways to appeal an overpayment determination.

Nothing in this column is meant to be legal advice about your specific situation. If you have questions, please contact the CF Legal Information Hotline at: 1-800-622-0385 (or e-mail CFLegal@cff.org). Providing 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.

Question

I received a notice from Social Security saying that I have been overpaid and that I owe SSA $10,000. I know I worked more than I should have a few years ago, but can they now make me pay back the money or is there a time limit for SSA to figure out that I was overpaid?

Answers

A. What is an Overpayment?

An SSA overpayment occurs when Social Security pays a person more than she should have been paid under their rules. For example: a person who receives Social Security Disability Insurance (SSDI) benefits can work only part time, making less than $1,000 a month from work activity. There is a one-time, nine month period where a person can make more than $1,000 a month. Once there have been nine months when a person has made more than $1,000 a month, SSA will review the person's file; then there is a risk of having benefits terminated, because SSA feels the individual can work making more than the allowable amount. Therefore, it is risky to make more than the $1,000 allowed under what is called the "nine month trial work period". After the nine month period, a person can make only less than $1,000 a month from work activity and still receive SSDI benefits. What usually happens is that a person may receive a bonus or work a few extra hours in a month, and that results in the person making over the $1,000 amount allowed in a specific month.

BETH SUFIAN

Social Security has no time limit within which it needs to determine that a person has worked too much and, therefore, was overpaid benefits. In addition, once a person makes too much money in one month, all months after that are considered months where the person has been overpaid, since benefits should have stopped in the month the person went over the allowable amount. Social Security may not figure out that a person has worked too much and, therefore, is ineligible for benefits until two or three years later. This results in a person receiving a notice of overpayment for a large amount of money such as $10,000 or $20,000.

An overpayment occurs when Social Security pays a person more than he should have been paid. If this happens, Social Security will figure it out at some point in time. Social Security reviews IRS records for each person receiving benefits on an ongoing basis. If earnings from work activity have been reported to the IRS, Social

Security will be able to determine if a person has received more than the allowable monthly amount from work activity. Sometimes it can take one to two years before SSA determines there has been an overpayment. There is no time limit in which they must uncover information about the date a person has made more money from work activity than is allowed. In the case of SSI, a person who has received money from any source that puts the SSI recipient over the resource limit will have an overpayment. As I have written about in prior columns, SSI recipients are allowed to receive up to $2000 a year from participation in research studies. If you need more information about this exception, please call the CF Legal Information Hotline. It is extremely important that you understand the work income rules for SSDI and SSI and that you do not go over the allowable amount.

The SSA notice of overpayment will explain why a person has been overpaid, repayment options, and appeal and waiver rights. A person should read the overpayment notice carefully and make sure he understands the appeal and waiver rights.

B. Appeal

If a person does not agree that there has been an overpayment, or believes the amount of the overpayment is incorrect, she can appeal by filing form SSA-561, which is available on line at www.ssa.gov or by visiting a local SSA office. An appeal must be made in writing and must be filed within 60 days after the person receives the notice of overpayment. SSA assumes a person receives the notice within five days of the date on the letter. An appeal should explain why an individual thinks she has not been overpaid or why she thinks the amount is not correct. Stating that it has been one or two years since the overpayment occurred will not result in the overpayment being waived. The time it has taken for SSA to notify a person of an overpayment makes no difference. This rule is being repeated for emphasis because many people think arguing about when they received notice of the overpayment will invalidate the overpayment. This simply is untrue.

C. Waiver

If a person believes he should not have to pay the money back, he can request that SSA waive collection of the overpayment. A person must submit form SSA-632, which can be found online at www.ssa.gov or by calling the CF Legal Information Hotline, or visiting a local SSA office.

There is no time limit for filing a waiver request. The sooner a request is filed, the sooner a person will have a decision on the matter. If an appeal or a request waiver is not filed, SSA will attempt to collect the overpayment by either stopping all benefits or reducing benefits and/or garnishing income tax refunds owed to the recipient. In order to have a request for waiver of the overpayment approved, a person has to prove:

1. The overpayment was not your fault; and
2. Paying the amount of the overpayment back to SSA would cause the individual financial hardship or be unfair for some other reason.

SSA can ask a person requesting a waiver to show proof of monthly income and expenses to verify the person does not have the funds to pay back the overpayment to SSA. SSA will stop recovering the overpayment only if a request for an appeal or a waiver is filed. While the appeal or waiver request is pending, SSA will suspend efforts to recover the overpayment.

D. Options for Repayment

If a person agrees that she has been paid too much and that the overpayment amount is correct, she will have repayment options:

1. A person receiving SSDI benefits will have the full amount of his benefit withheld each month, unless he asks for a lesser withholding amount and the request is approved by SSA. Full withholding starts 30 days after SSA notifies a person of the overpayment.

2. A person who receives Supplemental Security Income (SSI) will have 10 percent of the maximum federal benefit rate withheld each month. If she can not afford to have this amount withheld, she may ask that less be taken from the monthly benefit. Deductions from an SSI benefit check do not start until at least 60 days after SSA notifies a person of the overpayment.

3. A person who receives an overpayment notice but no longer receives SSI benefits, and instead receives SSDI, can request that up to 10 percent of her monthly Social Security (SS) benefit be withheld to repay the SSI overpayment.

4. A person may have stopped receiving all SS benefits because he has gone back to full-time work or has married a spouse whose income puts the SSI recipient over the SSI income resources amount. In these situations, the individual should either send a check to SSA for the overpayment amount or contact SSA to set up a payment plan.

5. A person who has stopped receiving SSDI or SSI benefits and does not pay the amount back to SSA can have the overpayment recovered from her federal income tax refund or from wages, if that person is working. Also, SSA can recover overpayments from future SSI or SSDI benefits or SSA retirement benefits. SSA also will report the delinquency to credit bureaus. ▲

Beth is 46 and has CF. She is a Director of USACFA. Her contact information is on page 2. She is an attorney who specializes in disability claims. You may send CF-related questions of a legal nature to: bsufian@usacfa.org.

SPEEDING PAST 50...

You're Too Old To Have CF!

By Kathy Russell

When I was diagnosed with CF, at age 12 in 1956, the medical people couldn't believe that I had CF because I was *too old* to have it. They were taught that people who had CF didn't survive to school age. I guess I knocked that theory into a cocked hat. My own private physicians could not believe that I had CF and they continued to say that I had a "fibrocystic lung condition". They just couldn't get their heads around the idea that maybe not all people with CF would die as children. I am glad that they were mistaken.

I was in my late 20s or early 30s when I stopped apologizing for being alive. By that I mean that when a medical person would learn my age and say, "You should be dead!" I would explain to them how rude that was and that they should read up more on CF. That is not to say that I have never felt guilt that I have lived when so many others have died. Having medical personnel act so astonished, that I was whatever age I was at the time, only increased those guilt feelings. Even at my age, I still have to remind myself that it is okay to live long with CF.

One doctor, on learning that I had CF, said that he had great success at disproving incorrect diagnoses of CF. I told him that I would be happy if he could disprove my diagnosis, but I would like him to tell me why I had the symptoms. After he looked at my chart, he said that there never had been any doubt of my diagnosis and he was amazed that I had done so well. (So much for getting the diagnosis overturned.)

I remember all too well how many times I would tell my CF docs about a particular problem and they would say, "Oh, Kathy, that's just you. It has nothing to do with CF." Among others, these problems included such things as chronic sinus disease, arthritis-like joint aches and pains, and the irritation and burning in my gastrointestinal tract that we now know as gastro-esophageal reflux disease or GERD, all of which have proved to be part of CF. While I am happy that the knowledge of what CF is has improved in my lifetime, I am sorry that all those problems may be in the future for young people who have CF. Of course, treatments for these various problems have definitely improved, so maybe they will have an easier time of it.

I was very fortunate to have parents who raised me to live. They taught me to live well and take care of myself. I made the decision to leave my pediatrician and start seeing an internist when I was about 16. That was an excellent decision for me. I was able to learn how to take care of myself while I still was at home and had my parents for backup. By the time I left home for further education, I was comfortable with making my own appointments, ordering (and paying for) my own prescriptions and doing all my treatments, without needing someone to nag me.

KATHY RUSSELL

I am so happy that the CFF has made the effort to create clinics for adults who have CF. It really was not good to have to go to a pediatric clinic with little chairs, kid's toys and little kids in the waiting rooms. When one is an adult, having to go to a pediatrician is not ideal. It is hard enough to make the transition from child to adult without having to go to a "kid's" doctor. After all, we are adults, not just large children. Thanks, again, to the CFF for making the effort to accommodate us. Our numbers will continue to increase, as more of us live longer lives.

One reason that it is difficult for pediatricians to treat adults who have CF is that we have adult needs. Most pediatricians are not equipped to deal with the problems of people who are aging. This starts with adolescents who are experiencing hormonal changes and romance for the first time, planning for college and careers, and looking forward to marriage and families. It goes right on to elderly people like me who are dealing with retirement, Medicare and old age.

As we age, we start having more problems with our digestive systems, incontinence, aching and stiffening joints, and all of the problems that all people face as they age. Some who may have had problems with loose and bulky stools start to experience constipation. Then they have to search for the proper treatment to alleviate that. (Oh, goody! Another pill to take or liquid to drink.)

No matter how many Kegel exercises we do, nothing ever will prevent

incontinence with a hard CF cough, when one is worn out from coughing. After the fifth or twelfth or seventeenth cough, our muscles just cannot hold any longer. Everything just gives up and cries, "Uncle!" Emptying one's bladder a little more frequently than normal can help, but nothing will totally alleviate the problem.

As we age, most of us will start to experience muscle, bone and joint pains. Some of us will have CF-related arthritis. Whatever forms our aches and pains take, they can be quite debilitating. I have arthritis as well as compression fractures in my spine. To say that I have a chronic backache is to be a master of understatement. My hands give me a fair amount of trouble, too. I find that I cannot write more than a few lines before my hand just doesn't work right. Any of you who have received anything handwritten from me know what I mean. I do not take any medicines for pain, because of intolerance to many of them. I have learned to live with it.

There is one really great thing about being as old as I am. That is that most of my friends are now starting to experience various health issues and I no longer am the "odd man out" when it comes to health. When I was young and all of my friends were healthy, I felt that I stuck out like a sore thumb. Now, I am just one of the geezers, and we all commiserate about our health, medical insurance and energy levels. Don't get me wrong, I would never wish ill health on anyone. It just happens with aging and, at long last, I am "normal".

The first third of my life was filled with education, starting a career, marrying, establishing a home and enjoying youth. The middle third was filled with hard work and attaining certain levels of comfort and pleasure. This final third of my life is all about enjoying life. We have wonderful family and longtime friends whom we love and with whom we love to spend time. We have known each other long enough that we all can be perfectly candid about how we feel and what we want to do. There are no competitions for place or prestige within our group of friends; it is a very comfortable place to be.

As you can see from what I have written, adults who have CF would be difficult patients for pediatricians to treat. They could treat parts of the disease, but they would have trouble dealing with many of the issues that we face every day. Pulmonologists who specialize in treating adults have a much better chance of treating us well and properly. (Thanks, Dr. Allada.)

I hope that all who care for adults who have CF will realize that CF, rather than being a disease of children, is a disease of people…some of whom die as children. With research into new medicines and treatments and with better care, there soon will be many more adults who have CF. This is something to celebrate and, believe me, I do celebrate it and am very grateful. Hooray for all of us adults who have CF! ▲

Stay healthy and happy,
Kathy

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

Mailbox

Hi...I would just like to comment that I have never had much opportunity to get to know others with CF, but *CF Roundtable* has been a huge help to me during my life of living with CF. In those articles, I learn about the disease, and make many "friends" who are sharing what I myself have gone through. That is hugely comforting. It is also informative. I have learned many things that I have spoken to my doctor about to improve my care. It is nice to know we are not alone in our experiences.

Nancy Johnson
Denver, CO

I have to share that I am a huge fan of *CF Roundtable*. It is a wonderful newsletter. Thank you to the entire staff for pulling such informative and moving pieces together.

Kathryn A. Sabadosa, MPH
Research Associate
The Dartmouth Institute for Health Policy and Clinical Practice
Lebanon, NH

Thank you. Your publication is always full of valuable information and inspiration!

Nora Furey
Malverne, NY

Thanks for *CF Roundtable*. Keep up the good work.

Kathy Yoder
Portland, OR

COUGHING WITH A SMILE...

Who I Am

NEW COLUMN

By Jennifer Hale

Hello! I am honored to be writing for *CF Roundtable* and I look forward to getting to know our readers. Here is a brief introduction of who I am, and then we can get to the topic at hand: "CF, It's Not Just For Children". I am 39 years old and was diagnosed at age two. I had lung issues that were thought to be allergies, but a sweat test told my parents otherwise. They were told that I had cystic fibrosis (CF) and the prognosis was not good. They were told to not expect their daughter to attend high school. That was in October 1974.

I am happy to say that October did not have to always be a dark reminder of CF. October was the month I married my wonderful husband! We now are able to replace a dark moment of a terminal diagnosis with the moment of a blessed marriage.

I got through high school and college, all at a standard pace, and dealt with the ups and downs of hospitalizations and sicknesses with a smile on my face. I had very supportive parents and still do to this day! As I have gotten older, CF has presented itself with a lot more complications in terms of CFRD, decreasing lung function, bowel issues, and whatever else that I can blame on CF, right?! HA! HA!

My daily regimen is full of nebulized drugs (three times-a-day), flutter/vest, enzymes, insulin, blood glucose checks, exercise, lots of sleep and good nutrition. I was in the "rat race" out in the corporate world but could not stay healthy and finally came to terms that a disability retirement would be the right choice for me. I am "retired" and living in Florida. Wait, I am only 39, not 79 – LOL! I am enjoying Florida and just looking out at the beach and the open water really speaks to my soul.

I named my column "Coughing With A Smile" because I always try to look at the bright side of the situation. Also, as many people with CF experience in their own lives, a CF cough is distinctive and my husband always knows where I am when we are out in public. He follows my cough; it is his yellow brick road to me! Therefore, "Coughing With A Smile" seemed to be an appropriate name for my column.

Now, for the topic at hand. "CF, It's Not Just For Children". I *love* this topic!! The reason it strikes such a chord with me is that it is *very* hard for people to realize and comprehend the seriousness of living and breathing with adult CF. If I can count how many times I have been told, "You do not look like anything is wrong with you," I could have retired to Florida a long time ago! On a positive note, I am happy that this is a topic of discussion to begin with in the CF community. To be an adult with a "terminal" disease and see all the progress that has been made since I was diagnosed has been nothing short of amazing. Since CF is an orphan disease, I am so grateful for the advances that have come throughout the years in terms of the drugs, medical care and research. Living with CF as an adult now presents a lot of different obstacles.

JENNIFER HALE

Being an adult with CF has been very different from how things were when I was a child. As a child I was much more resilient in terms of my CF. My hospital stays were a chance to see my favorite nurses and to decorate my IV pole. As an adult I like to stay far away from the hospital, opting for home health care instead and explaining to people that the undecorated wrap around my arm is protecting my IV PICC line.

Being an adult means having to rely more on yourself to get through all the meds, therapies, tune-ups etc. It is hard to be your own cheerleader and motivator all the time! Lucky for me, my husband is a huge help and my parents still are in my corner!

When I was a young adult, my life was all about school and how I could get through college while being around people who might be sick in the classrooms. I had to maintain a social life and still be compliant with my care. I was able to stay compliant and get through college in only four years! At the same time, I did not do as many nebs as I do now. I did not have CFRD and my lung function was a lot higher than it is now. I am thankful I had increased lung func-

tion while getting through school and I was able to enjoy college life.

As an adult going into the corporate world, I had more control about avoiding "sick" people, or so I thought. I once again found it hard to stay away from people who had colds – since everyone went to work sick – and still does. It is the society we live in that, unfortunately, dictates there is no rest for the weary. I found that my own health was declining and I was not able to work in the "rat race". That is when I decided I needed to get on disability. What a hard decision that was to accept.

In your early 30s you are either working or working to have a family, and I was now doing neither! Disability was hard to accept, which is a whole other topic of discussion! Now my "JOB" is me and taking care of myself so I can stay as healthy as possible. The problem is that I cannot fire myself! I have to keep going and doing all the meds and therapies. My advancement is not promotions, raises or a different job description. My promotion is being able to get through the night without sleeping on five pillows, getting out to the gym on a regular basis, ordering meds and supplies and doing all my therapies and medications on schedule.

Being an adult is also realizing what costs are associated with having CF. Whoa! Do the bills add up or what? Dealing with insurance companies is as frustrating as a bad itch on your back that you can not reach. Now, not only am I an adult dealing with a terminal disease but also handling all the paperwork, insurance companies, doctor appointments and proper management of therapies and medications, too. I can go on and on; wait, maybe this is a JOB. I am the CEO of me!

Having CF as an adult means you also have to deal with the decision to have children or not. That was a big decision for me and to think I will never be called mommy, sometimes really pulls at my heart strings. My husband and I decided my health was too important and high maintenance, and to have a child would be too hard for us. That was our decision for us and what was right for our lives.

Now, it is just me and CF which is kind of like a baby. It does require lots of care, daily feedings and tons of attention! CF is not just for kids, and being an adult with CF presents a new set of complications and emotions. I sometimes wish the sympathy and empathy a child receives for dealing with a disease that is so horrendous would carry on into the adult life of someone with CF. The people who meet me can not "see" the disease and people just do not always have that same compassion for an adult with a "hidden" disease that they would have with a child. But there are people I meet who do "get it", and that makes up for the trials and tribulations of dealing with CF as an adult. I am glad we can talk about CF adults and I am happy to have these "adult" problems to deal with in my life. CF is challenging and a rough road to walk. To have that chance to become an adult with CF is a wonderful opportunity. Don't take a day for granted and, as I always say, "Keep up the good fight; it is worth it!"

Until next time, my friends! ▲

Jennifer is 39 and has CF. She lives with her husband, Mark, in St. Petersburg, FL. You may contact her at: jhale@usacfa.org.

Benefactors

BRONZE

Susan Le Boeuf
James Chlebda
Mary "Toddy" Coon
Eileen Gallagher
Gay Kane Lazur
Kim Nunnari
Lisa Nunnari
Brian Weinstein

SILVER

John & Nancy Stanley (in memory of their son, John Alan Stanley)

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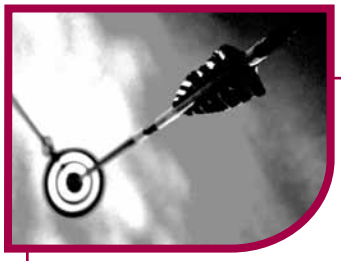


FOCUS TOPIC

CF, IT'S NOT JUST FOR CHILDREN

Staying Optimistic

By Maria Fioccola

Frustration settles in about hour 72. Hour 100 arrives and you find the strength to move forward. No matter how many beatings you've had, you are scheduled for more. Popping pills, eating the days away and hydration are of only some importance compared to the results of PFTs, blood tests and more.

You meet very nice nurses and doctors, all of whom seem to be knowledgeable. Yet, you aren't getting any answers. You feel support from family. Yet, they don't understand. Friends are few. It seems the word "hospital" scares people. To me, it's my booster club; it's "Club Med". It could be worse and, yes, Club Med isn't all joy by any means. However, I know and feel that this is where I need to be. The things many take for granted, I try not to. "It's about the moments that take your breath away." I people watch. I socialize. I learn my surroundings. I learn schedules and each step gets a little easier with faith. I wait. I breathe.

A simple tickle in my throat causes chaos inside. My lungs jarred, my face red, I cough. When I laugh, I cough and, in a twisted way, I think about the Mucinex® commercial. Yes, the little mucus guys. They like to build in my system, they don't let me know they are rooming and they start a party. I'm the party pooper. I giggle once again, as I know you are grinning, yet hoping this battle will be won. Nine days and five pounds later, I am released from this place. This place where no cure has yet been found leaves an imprint in my body – I know they do everything they can to help.

In my experience, I never was a child with cystic fibrosis (CF). I lived in a household where doctors were frowned upon and tests were "false". I lived in a household with a cloud of smoke. I find myself very lucky to be alive.

I always was a happy child. I had a close-knit group of friends, many of whom didn't know of my CF. I was always the tiniest in my class, yet the oldest with a birthday in September. I grew up in Melrose Park, IL and attended a Catholic school from kindergarten through eighth grade. We had uniforms and small classrooms.

> "In my experience, I never was a child with cystic fibrosis (CF). I lived in a household where doctors were frowned upon and tests were "false"."

I remember back in kindergarten my primary physician tried to explain CF to my mother. It was the year that I was put on Pancrease® MT tablets. The doctor explained to my father that it was like me eating a piece of bread to absorb nutrients and help me to gain weight. In the sixth grade, while other classmates were hitting their growth spurts, I was not. I decided after all the teasing that I didn't need my medication at lunchtime and started hiding my pills.

It wasn't until the eighth grade that my primary physician found out my secret. I was then put back on my medication with still little information of what CF was. That year in science class we read an article about it. To me, I wasn't that patient. I wasn't sick. I wasn't in the hospital at any point during grade school. I was about 80 pounds that year and ready to start high school in a new area. After I graduated eighth grade, my family and I packed up and moved to Lindenhurst, IL. I was up for the challenge of a public school; new people, switching classes, the bus system and best of all NO UNIFORMS! It couldn't be that hard, right?

First day of high school came pretty quick. I was nervous. Over the summer, I had gone to a "new student orientation" to learn where my locker was and what my class schedule would be. I had trouble with my locker and became very tired walking from class to class. At this point, I thought it would be good to research CF.

My father, still in denial, had told me that there was no need to read about it. He told me the doctors were wrong. I'm sure this is a hard disease on parents. However, I needed to know who I was. I felt lost in my own body when I started reading about treatments, medications and life expectancy. Life expectancy of 37 years was very scary to think of. However, I still didn't feel sick.

Sophomore year rolled around and I just wasn't feeling right. My stomach was in knots most days. I chalked it up to lunch room food. After all, I didn't get a stomach ache until after lunch. It made complete sense. I went to the nurse's office

almost every day for two weeks. They would call my mother, put a cold cloth on my head and sit me down for a while, before I was ready to go back to class. I was also in Drivers Education that year. Driving made me nervous so, maybe, it was nerves bothering me.

It was October 22 when I went to the nurse's office with a stabbing pain in my gut. I knew something was wrong, but since I had been going to the nurse's office often after lunch, we went through the same routine. I remember telling her that today was different. I was doubled over in striking pain. I had tears in my eyes and felt I was going to lose my lunch. I wanted my mom. The nurse had the chance to call my mom. My mom showed up and took me out to the car, where I lost my lunch. Normally, after something like this, most people tend to feel better. I went home and lay on the couch hoping that this feeling wouldn't come back. An hour later I was doubled over again.

At about 4 pm, we went to the ER. There I found out I had ten gallstones and needed to have my gallbladder removed as soon as possible. Yuck! Surgery! I had never been hospitalized and definitely knew nothing about surgery. I was scared but deep down knew I would be okay. I prepared like a champ. I did everything the doctors wanted and tried not to worry about all the school work I was going to miss. After surgery, I was released the day before Halloween. I was told that I shouldn't return to school for another week, making it two weeks of missed classes.

Once I was "normal" again, I felt I had to continue my search about CF. I did a school report on it as well. As a junior now, I knew I had to start making decisions about college, another chapter that I didn't know much about. Neither of my parents went to college and I was too young to remember when my older brother started. I signed myself up for early

> "A simple tickle in my throat causes chaos inside. My lungs jarred, my face red, I cough."

graduation that year. I was looking forward to getting out of high school in January 2003, instead of June, though I would come back to walk down the aisle with my class.

Oh, boy. Yes, high school wasn't only about school. My growth spurt with full-fledged hormones seemed to come overnight. Freshman year, I laid my eyes on a very special boy who lived in our subdivision and was on my school bus. I remember telling friends and this boy that I had CF. I also remember that most did not know about it and reiterated that I didn't look sick. That boy had asked me out and that was the beginning of the "I can do anything" phase.

Throughout high school, many people came in and out of my life. This boy had moved away as well, though we kept in touch. He became a best friend to me. Senior year was a chance for me to move on, for me to walk into another school with nobody knowing I had CF. Of course, by this time I was managing my tablets well and gained some weight. I was now above 103 pounds. This weight didn't stick. I was 93 pounds in no time, with the stress of the new environment.

I feel I was forced to grow up. My mother became pregnant when I was nine years old and my older brother was already 18. He was out of the house most the time, and I became a great big sister to my new little brother. I helped my mom feed, change, bathe and put my little brother to sleep on a daily basis. My mom would thank me all the time for being so helpful, but I would always dislike cleaning my room. Once I reached my senior year, I felt my little brother looked up to me as I looked up to our older brother.

Ah, adulthood. Knowing that the life span of CF patients was a young 37 years, I thought this would be my time to start living life to the fullest. Between 18 and 22 years of age, I accomplished many hurdles. I tested for my license and started college. I had a liver biopsy and was put on medication for cirrhosis. I married and divorced young. At age 22, I started focusing on me again, not what others wanted me to be or do. I researched CF Adult Clinics. I fought for disability and won. I received my own insurance through the state.

By age 23, I began at Northwestern Memorial Adult Cystic Fibrosis Clinic. I started my CF journey not knowing anything. I felt like a child beginning school again. I felt I needed more support, motivation and care than I would ever need as an adult. I was introduced to new doctors, medications and treatments that I once had read about. I was learning why I didn't feel sick or look sick. I learned that these medications and treatments would help me beat the odds of living past 37 years of age.

My life has taken many twists and turns. CF lives with me, I don't live with CF. I am now 27 years old and feel optimistic that I will survive past 37 years. ▲

Maria is 27 and has CF. She lives in Gurnee, IL. She has a very supportive boyfriend, who has two boys, with whom she enjoys spending time. She enjoys writing - from short stories to poetry, and enjoys listening to all sorts of music - from country to today's hits. Music inspires her mood and her mood inspires her to write. She is very social and loves meeting new people.

CF, It's Definitely Not Just For Children

Our lives with cystic fibrosis are unique, yet similar. Our CF community continues to grow with early diagnosis of infants and young children as well as delayed, often elusive, adult diagnoses. Here, three women share their own stories of being diagnosed with CF in middle age. What is your story?

Cystic Fibrosis? Thank goodness!

By Laura Mentch

> "I challenged myself to get better on my own - until I would, finally, drag myself into the doctor's office for antibiotics, year-after-year-after-year."

"What did I do to cause you to be so sick?" my mother asked the last time we were together. I was 49 years old. My health problems were always a challenge for her. Together we went to so very many doctors' appointments – the pediatrician for sinus, ear and lung infections, the allergist for testing and shots twice a week, followed by ice cream at Carvel's, and, when I was very young, the doctor with the mirror on his head. I had an air filter in my (rug-less) bedroom, home-made wheat-free bread, lots of allergy pills, nose plugs for swimming and the "K" machine, a nasal aspirator hooked up to the kitchen sink.

The older kids called me "Goo". My mom always wanted to see what was in my tissue. When I could not breathe at night, we sat in the bathroom with a hot shower running. We did all these things for my "allergies". This was my childhood and my mother's burden.

I left her home for college with serum for my shots and an antiallergic cover on my pillow. Bronchitis each spring and fall was routine. With each new infection I challenged myself to get better on my own - until I would, finally, drag myself into the doctor's office for antibiotics, year-after-year-after-year.

My mother died a week after our last visit. While planning her memorial service I became really sick. That early summer episode began months of persistent lung infection and much Cipro and Levaquin. I was still coughing in March and asked my doctor, "What's in there?" My very first sputum culture showed Pseudomonas aeruginosa. My doctor wondered about that and referred me to a (new) pulmonologist. I had my diagnosis of CF by my 50th birthday.

LAURA MENTCH PRESENT DAY AND, ABOVE, IN 1954.

Today I eagerly walk into the CF clinic and am beyond grateful for the doctors and nurses who know how to help me and teach me how to take care of myself. I believe my mom would be relieved and thankful to finally have the answer. ▲

Laura is 58 and has CF. She lives in Bozeman, MT with her husband of 35 years, Michael Brody. They are the parents of three adult children and two dogs, still at home. Her work in health education focuses on sexual and reproductive health and she offers her knowledge and experience to the CF community. She can be reached by e-mail at: laura21@bresnan.net.

Triple Mutant

By Juliet Page

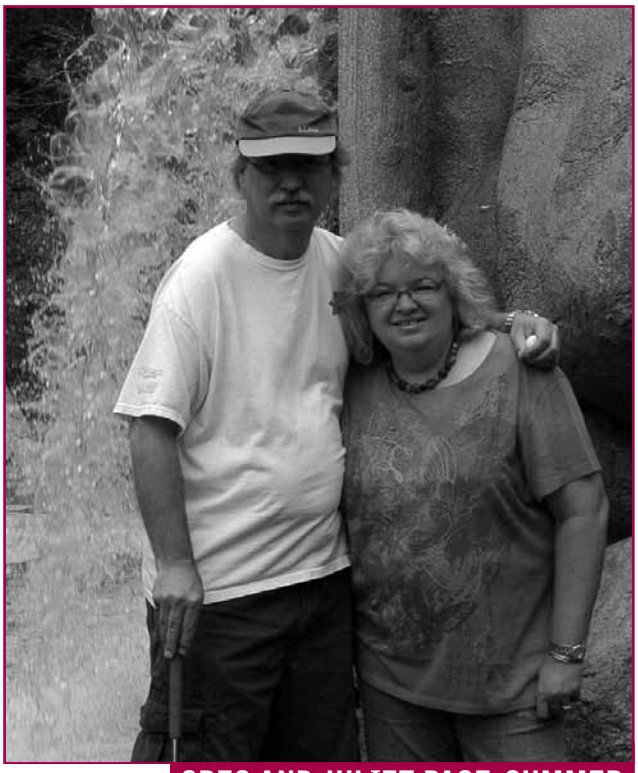
I was diagnosed with CF just over three years ago, at age 43, via genetic testing. I've had unrecognized CF symptoms all my life: chronic cough, frequent bronchitis and lung infections, and some mild GI issues. When I was a kid, they tested me for all sorts of things but never came up with the CF diagnosis, primarily because I'm pancreatic sufficient. Somewhere around 2005, I caught my first mycobacterial infection (MAC), which left me with reduced lung function. I decided it was time to revisit the 'no diagnosis' of my childhood, given the 25+ years of medical advancements since then. Fortunately, I had a very cooperative GP, who was equally determined to "figure this out", and who ruled out many possibilities and also referred me to several specialists.

My sweat test was in the "inconclusive" range (just like when I was in High School). My doctor sent me to the Adult CF Clinic at Johns Hopkins in Baltimore, because he thought some of the newer CF treatments and medicines might be of benefit to me. Hopkins suggested a genetic test to "rule out"' CF.

In March 2008, I got that unexpected, yet life-changing, call from Dr. Sosnay. Bingo! Positive for CF. My results were not just positive, I had three identified mutations. Two are on one chromosome, and one on the other. What a polar shift in my line of thinking and attitude.

I was relieved to finally know the root cause of my cough and lung issues. All my seemingly random issues over the years likely were related to CF. It all made sense. I've been very lucky that, until recently, my CF never really slowed me down. I finished high school (despite being sick a lot), went away to college, got married, attended grad school while working full time, had a daughter and a great engineering career without ever knowing about CF.

> "I was relieved to finally know the root cause of my cough and lung issues. All my seemingly random issues over the years likely were related to CF."



GREG AND JULIET PAGE, SUMMER VACATION IN CAPE COD IN 2011.

In retrospect I'm glad I didn't know, because things might not have turned out the same. Adjusting to the demands of advancing CF has not been easy on me or my family, but I'm grateful I am receiving the best care and guidance possible. The CF team at Hopkins has graciously answered my endless stream of questions, even those for which there are no answers… yet! I'm especially thankful to have found wonderful online support from other adults with CF, who have helped me immensely with the CF learning curve! ▲

Juliet is 46 and has CF. She and her husband, Greg, and their 16-year-old daughter, Alexanna, live in the Annapolis, MD area. Juliet works full-time as an acoustical engineer at Wyle Labs and is head of the Environmental and Energy Research Group. She is a Master Watershed Steward for Anne Arundel County, volunteering with environmental restoration and preservation of the Chesapeake Bay. She also is the patient representative for the CFTR2 Genetics Project at Johns Hopkins. Her hobbies include glass bead making (lampworking), silver work and jewelry making, restoring a vintage Corvette and power boating and sailing on the Bay – and about a zillion other things. Oh, for more hours in the day! You may e-mail her at: Juliet@julietpage.com.

A Marathon Walk to Cystic Fibrosis

By Kathleen Wilson

> "Now nine years later... I truly can say, 'Yes, I am getting older, but life is better.'"

When I turned 50, I decided to walk (can't run) a marathon to show that, "I'm not getting older, I'm getting better". My husband, Art, joined me in this endeavor (great couple time) and we joined Leukemia and Lymphoma Society's Team in Training program for the six-month training and race in Denver, Colorado.

About half way into the training I caught a cold and struggled with a cough that wouldn't go away. My primary care physician sent me to a lung specialist, just weeks before the race. Nothing jumped out as a cause for the persistent cough, but he prescribed an Albuterol inhaler for the race. And, yes, with the help of the inhaler and a gallant husband (he gave me his dry socks and wore my wet ones) we completed the 26.2 mile race in seven-and-a-half hours on a cold rainy October day in Colorado.

Eighteen months later, after a bronchoscopy, many CT scans and an open lung biopsy, the lung specialist gave up and sent me to Dr. Fontenot, a specialist in the pulmonary critical care division of National Jewish Health. He routinely runs the basic genetic tests for CF and, within a few weeks, I was diagnosed with CF.

KATHLEEN WILSON

Now nine years later, thanks to Dr. Nick and the great team at National Jewish Health, I truly can say, "Yes, I am getting older, but life is better." ▲

Kathleen is 61 and has CF. She and her husband of almost 25 years, Art, live in Castle Rock, CO. They have two grown children, Robin and Mark, and four grandchildren. She is a retired Montessori Pre-School/Kindergarten teacher. Her hobbies include stone carving with techniques and stones from Zimbabwe, watercolor painting, and fishing. In addition, everybody at Starbucks knows her well. She also is a Discalced Carmelite of the Roman Catholic faith for over 10 years. She credits her faith and a loving husband for her good attitude about life in general and CF specifically. You may e-mail her at: kathleenllwilso@yahoo.com

Hidden Agenda

PHOTO BY ROBERT WRIGHT

The invisible motivator

I'm standing in front of fifty architects and engineers
I'm telling the way I want it to be
So we help sustain mankind and our modern lifestyle.
So we'll stop blowing up mountains to find coal.
So we'll prevent climate change progressing.
But it all started with air quality
Me discovering the giant tailpipes in the sky,
spewing coal ash in my air so we can feel cozy indoors.
My audience can't see my core motivation,
my lungs struggling against the bacterial cesspool destroying them from inside.
These lungs crave fresh air, a deep unrestricted breath, healing.
Something these lungs will never experience again.
But their malfunction is your gain, they push me to fight for cleaner air.
The hope our successors will read about our short-sighted lifestyles in history books.
This is my fantastical vision, and these crippled diseased lungs that you don't see,
are really what motivated me, to want to feel like part of the solution.

-E. Hyman, 2010

"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...

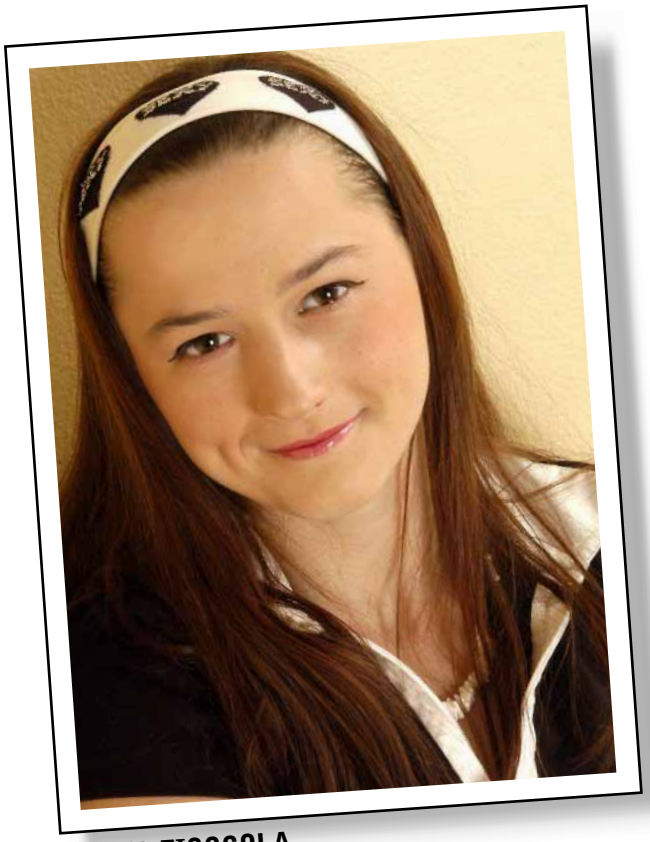
ANNA MODLIN, LINDA SHAK, USACFA DIRECTOR MAGGIE SHEEHAN AND HER FIANCÉ, THOMAS WILLIAMSON, AT THE 2011 CFRI FAMILY EDUCATIONAL CONFERENCE.

BETH SUFIAN CELEBRATES HER 46TH BIRTHDAY WITH DAUGHTER, ISABELLA, AND HUSBAND, JIM PASSAMANO.

EXTRAORDINARY TIMES AT THE 2011 CFRI TEEN & ADULT RETREAT.

KATHY RUSSELL AND HER INOGEN G2 ON THE RUSSELL'S 1954 CASE TRACTOR.

ANDREA EISENMAN IN KILKENNY, IRELAND IN SEPTEMBER 2011.



MARIA FIOCCOLA

IN THE SPOTLIGHT

NEW COLUMN

With... Anabel Mariko Stenzel & Isabel Yuriko Stenzel Byrnes

By Jeanie Hanley and Andrea Eisenman

Welcome to our inaugural *In the Spotlight* column. We are fortunate to have not just one but two interviewees who have graciously agreed to answer our questions about their lives. They probably need no introduction as they are both very involved in the world of CF. They are creative forces to be reckoned with as authors, subjects in a film based on their book, frequent contributors to this publication and gold medalists at the US Transplant Games. And when not fighting the good fight, they also have fun playing bagpipes, hiking, climbing mountains, and relaxing with their husbands. Without further ado, here they are in their own words.

Age: We are 39 years old. Ana was born 2 minutes before Isa on 1/8/72.
Married? Yes, Isa to Andrew Byrnes 6/27/98; Ana to Trent Wallace 6/5/10
Kids?

Ana: My lungs are my kids and my gut is the evil twin. No desire for children. I also have a "no child left behind" policy.

Isa: I'm a bit too concerned about long-term survival to consider children. Thankfully my husband is not fond of kids either.

Diagnosed w/CF and how/why:

Ana: I was diagnosed with CF at three days of age due to meconium ileus (MI). Isa did not have MI. Because we were probably identical twins, she also was tested. Back then a sweat test was used and it was blatantly positive. In a way the MI was a blessing in disguise since my mom is Japanese, and I'm sure our diagnosis would have been overlooked if I didn't have that classic symptom.

Your alleles if you know them:

We are "triple mutants" which means we have one delta F508 from our German dad, and 2 mutations– R347H and D797A - on the other chromosome from our mom.

Has having a twin made it easier to have CF?

Ana: Absolutely! Being twins was the best blessing of our lives and we truly believe we wouldn't still be alive without each other. With our severe CF, twinship allowed a symbiotic support system since infancy. We shared our disease, encouraged and comforted each other; and the illness experience was made less lonely and isolating, even if there were times we fought like cats and dogs. Also, we received a lot of attention because of our "twin-ness". I think that helped us create an intense team dynamic from an early age. Our symbiosis allowed us to accomplish things we may not have been able to without each other, such as college, grad school, jobs, living abroad, writing a book, but most of all, surviving to almost 40. When one inhales, the other exhales… that is the beauty of our twinship: the power of two.

Isa: Ditto. We are very close, at the expense of feeling less connected to our parents. It's a miracle we are able to have "normal" relationships with men. But, normal is relative! Overall, we struggle with balancing our relationship: being completely transparent with each other about everything while respecting boundaries and very real differences in our personalities.

ANA STENZEL (LEFT), AND ISA STENZEL BYRNES IN FRONT OF THEIR MOVIE POSTER AT THE L.A. PREMIERE SCREENING OF THE "POWER OF TWO," AUGUST 2011.

The hardest part about being a twin has been comparing each other. I was always the healthier twin and felt guilty that Ana was sicker. We compared lung function, weight, athletics, etc. all of our lives. We had to remind each other that everyone with CF has their own course, even twins.

Another word about cross-infection: some families worry about siblings who both have CF. Ana and I did each other's chest percussion for 20 years, and we still cultured different strains of pseudomonas. I think our care for each other—physically and emotionally—far outweighed any risk of cross-infection that existed between us.

> *"God forbid, but if I needed a third transplant, I would take it in a heartbeat. It is the second best thing that ever happened to me (besides being born a twin). – Ana"*

Transplant and when/ how many years:

Ana: 6/14/00 bilateral lung transplant, rejected in 2007. Second bilateral lung transplant 7/13/07.

Isa: 2/06/04 bilateral lung transplant. No plans for second!!!

Where did you have your transplants?

Our transplants were by Dr. Bruce Reitz at Stanford Hospital.

For Ana: Why did you have to have another transplant? Can you elaborate?

Unfortunately, I went into rejection of my beloved first transplant in October 2006, and despite attempts to treat it, within eight months I was back on oxygen and in a wheelchair, feeling worse than I had ever felt with CF. I had another transplant because my doctors offered me one. It is rare to be in good enough shape to withstand another and I felt I was a superb candidate, due to my compulsive fitness, until rejection hit. I had just met a wonderful man (who later became my husband) and our book was on the verge of being published. I JUST WAS NOT DONE WITH LIFE YET. That's why I took the risk of another transplant. If I had died during the process, at least I had tried. I felt really guilty and struggled with being on the list again, since I knew people who had died waiting for their FIRST. My doctor convinced me the San Francisco Bay Area had the highest donation rate in the country and I wasn't "stealing" from another patient who needed it.

For Isa: Do you worry about needing another transplant?

When Ana had her first transplant, it drove me crazy when people would ask me when I was getting a transplant. Now it's the same for a second transplant. We are twins, but we have our own paths to live out. I can learn from Ana's lifestyle what may have played a role in her rejection, but I also know sometimes rejection just happens. I think it's normal to worry about rejection. I am very attached to my new lungs and easy breathing, and don't want to ever lose this gift. That's why I'm very focused on exercise and bag-piping to help my lungs stay healthy.

What you are able to do now that you could not do before transplant?

Ana: Fortunately my first transplant was the epitome of success. I had no diabetes, no major complications (except transient post-transplant depression afterwards), no sinus issues, and no gut issues for six years. I was never tired, was able to return to work, take up exercise to levels I had never imagined, traveled all over the world, hiked, backpacked, ran, swam, biked and was hospitalized only once – for a bike accident, ironically. Part of this luck was a combo of genetics, personal fitness, good medical compliance, good emotional state, and a wonderfully healthy CMV/ EBV-negative donor.

After my second transplant, I felt like I aged about 10 years – it kicked my ass. I developed diabetes, sinus and gut issues that I have to attend to more vigilantly and have been hospitalized 10 times for opportunistic infections like CMV, RSV and recurrent cellulitis skin infections. I am back to being a patient, after a six year vacation from that with my first transplant. I have no regrets however; my current health challenges are humbling. I am a deeper, more spiritual, accepting person and my health challenges make me slow down once in a while, which is a good thing (we all need a "control/alt/delete" now and then). I am fairly fit for a bilateral lung transplant recipient and I live quite a normal life. I am still able to hike, bike, jog, swim and backpack, but at a slower, easier pace.

God forbid, but if I needed a third transplant, I would take it in a heartbeat. It is the second best thing that ever happened to me (besides being born a twin).

Continued on page 20

Isa: Given that I was about 12 hours from expiring before my transplant, every day above ground is a good day. The little things I have to deal with are just that: little things that are the price to pay to still be here. I find writing about my medical history excruciatingly boring, so I'll refrain. I deal with normal post-transplant stuff but also recognize that I've been tremendously blessed to have seven very good years.

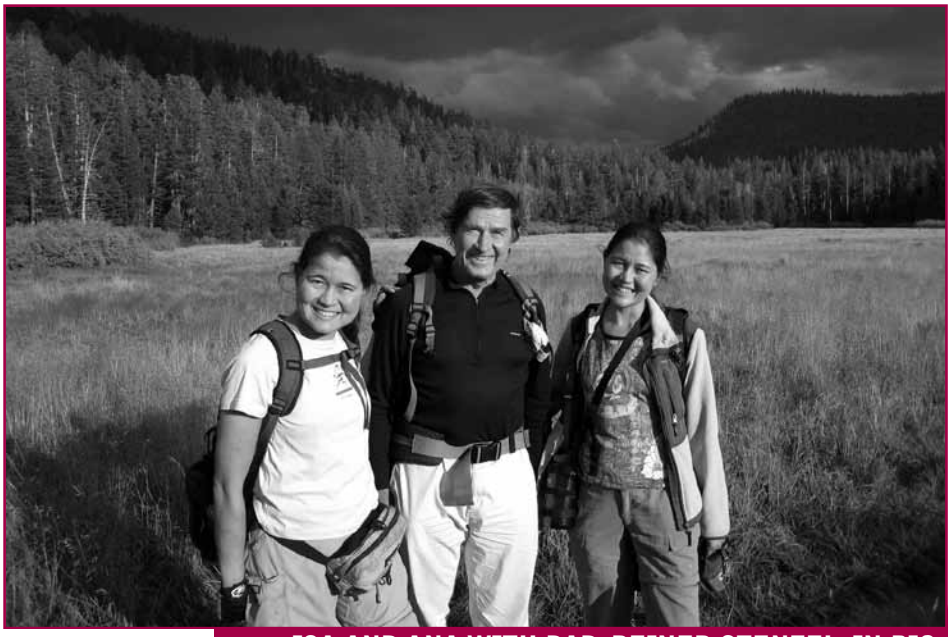
How did "The Power of Two: A Twin Triumph Over Cystic Fibrosis" come about?

Ana: My mother always encouraged us to write out our feelings, when we were hospitalized as kids. After over 35 hospital stays by the time we were 20, we had volumes of journals. She always dreamed of us publishing some of the content in a book.

When we were in our 30s and Isa was facing advanced CF, unable to work and on disability, she spearheaded moving forward with making this dream a reality. She really was the one who started taking writing classes, going to writing conferences and reading many other memoirs. We wanted to show what our generation has been through – where for some, CF transformed from a fatal disease to more of a chronic one. Based on memory, our childhood journals, photos, and our family's input, we created our book. Our focus was on presenting a different spin from other CF memoirs, by including our rather unique experiences growing up as tri-cultural twins of Japanese/German immigrant parents trying to make it in the United States with CF kids. With the help of friends-turned-editors, we created a draft that we pitched to an agent.

Having a book published has opened up so many doors and helped me do what I've always wanted to do, which is spread the word about CF and encourage others who are dealing with this crazy disease. Because of our book, we've had speaking opportunities and book signings all over the country, meeting so many amazing families in the CF and transplant world. That is by far the best part – growing our CF family.

Since writing the book what is (are) the most significant life event(s) that have occurred? Is another book in order?



ISA AND ANA WITH DAD, REINER STENZEL, IN BIG MEADOW, EL DORADO NATIONAL FOREST, CALIFORNIA.

Ana: Since publishing our book, the most significant life event is that we are still alive. As many know, life post-transplant is a gift and each year that we celebrate a transplant anniversary or birthday TOGETHER is a gift from God. Our lives have truthfully become busier and more stressful than ever before as we are traveling, speaking, making the documentary film inspired by the memoir, becoming more public globally, while still trying to work, maintain our homes, pay attention to our husbands and deal with the ups and downs of chronic health needs. My personal greatest life events since the book have been surviving a second transplant and going on book tours, getting married and being part of the making of the documentary film, "The Power of Two" (www.thepoweroftwomovie.com).

Not sure if another book is in order. I think my husband would divorce me if I did that; he already thinks I'm a workaholic. I'm just trying to go with the flow of the film's upcoming release and see where life will take us next, while trying to stay healthy.

Isa: In addition to still being alive, I think my most significant life event has been staying married and having my husband spearhead the documentary film inspired by our story. I'll be honest and say I'm pretty tired of hearing about my story, so I'm not likely to write another book. Instead, I enjoy occasionally blogging and writing for *CF Roundtable*. But the book has opened up other doors: I'm excited about leading writing groups at hospices and encouraging others to share their stories.

When is this movie, based on your

book, available to the public?

The movie will be shown at film festivals this summer and fall, and we're very excited to see what happens. The film will be available to non-profits, medical institutions, and interested organizations for screening and panel discussion as of November 1, 2011, and we encourage those interested to see: http://www.thepoweroftwomovie.com/community-screening-faq/.

What are you hoping people come away with from "The Power of Two" movie?

Ana: The main goal of the film is to raise awareness about CF and the miracle of breath through lung transplantation. Ultimately, I hope it inspires viewers to sign up to become organ donors, increase their own motivation to take care of their health, and live life to the fullest. I hope the cross-cultural perspectives and cultural barriers to healthcare access portrayed in the film will show Americans that compared to some countries, the USA has many positive aspects of our healthcare – the most important being advanced medicine like transplantation, patient advocacy, self responsibility and peer support.

Isa: I'm also hoping more people will support CF treatment and research. I want the film to show that having an illness is not always just a bad thing. Our lives have been incredibly enriched by our CF community. The film also features a number of other CF patients, and I want people to see that our story isn't that unique. I hope our story will inspire people to get involved in their illness communities as a way to find connection and meaning.

How important are the transplant games to you?

Ana: I've attended five USA transplant Games since 2002. To me the Games are a reward for transplant – they are a sacred place to bond with fellow transplantees, rejoice with fellow CFers at the miracle of being free from CF, and to push our bodies in new ways to do the best we can in athletics and give thanks to donor families. I have never been able to do sports before, so the Games opened up a new world for me. They are extremely important to motivate me to pay tribute to my donor, celebrate my gift of life, and exercise. Every time I am on that starting block about to jump into the pool, I ask myself, "Why the hell did I sign up for this?" and my answer is, "Because I can," and I do it for those peers who died before me who can't.

> *"Given that I was about 12 hours from expiring before my transplant, every day above ground is a good day."*
> *– Isa*

Isa: I attended the 2004, 2006, 2008 and 2010 Transplant Games. The point is not about winning, it's about challenging myself to be in the best shape I can be. It is a privilege to be an athlete!

What are your goals?

Ana: Menopause…And to live with my lung transplant as long as I lived with my pre-transplant CF (28 years each).

Isa: My goals are to stay healthy for as long as my fate allows; to grow old alongside Ana and many of my friends; to see our film live out its potential; to witness the "cure" for CF; and, on a personal level, to perfect my bag-piping skills; to hike some Sierra peaks and Machu Picchu; to teach writing for healing classes; to work in hospice or counseling regularly enough to obtain a license in clinical social work; to make a difference in the world that is bigger than, and outside of, myself.

What would you tell someone who has CF and is awaiting a transplant?

Ana: I'd tell them that patience is #1. It takes patience to wait for lungs but more patience to feel better after the surgery. I would tell them tips like what to pack for the hospital, how to disinfect the pseudomonas from your home post-transplant, and to "drink Miralax" when you get the call, if you are prone to blockages as I am, since the surgery just exacerbates that. I would have them write goals of what to do after transplant to have something to live for. I would encourage them to harness their spiritual, social, emotional and mental reserves and to build those blocks of resilience up high. I would tell them to hope for the best, but don't be disappointed for some UNEXPECTED bumps in the road that are so common and can be so individual. I would tell them that there are great resources like *CF Roundtable* filled with amazing people who can provide advice and support. I would remind them that no matter what happens, you, as a patient, are the one in charge. You know your body best, and you will be the only one who can truly communicate, "I think, I feel, I want" to those around you to reach your maximum quality of life post transplant.

Continued on page 27

WELLNESS

The Perfect Workout... At Any Age

By Julie Desch, MD

After living 50 plus years as a fitness nerd, I have a few ideas about what constitutes a perfect workout for me. Of course, this all depends on which "me" shows up to exercise that day. Is it the "feeling great" me, the "getting back up on my horse after a round of IVs" me, or the "not exactly sick, but not feeling at my peak" me? The right workout for each situation will be vastly different. In addition, the "me" who shows up at age 50 is most certainly not the "me" who showed up twenty years ago. Yet, the basic constituents of the hour or so at the gym are the same.

Is there a "perfect workout" for you? I think there is, but I guarantee it is unique to you, and unique to you on this particular day. From the 10,000 foot point of view, the "perfect workout" is the one that you will do, consistently, and if not enjoy, at least not abhor. It should leave you feeling tired in a good way, so that you know you did some work, but not so exhausted that you dread the next encounter with your inner athlete. As you know, life with CF waxes and wanes, so the perfect workout is very much a moving target. Some days, 20 minutes on the elliptical is the right amount, while other days, 5 minutes on the stationary bike is what your body needs. On really awesome days, a 5-mile hike in the woods fits the bill perfectly.

But whatever state your health is in, the components of each workout should be the same, modified to suit your body with its particular issues. Each component is important, and the order that you complete each component matters. I'm going to run briefly through each and offer some suggestions for specific exercises that work for me and might also benefit you.

The order is as follows:

1. soft tissue work
2. mobility exercises
3. corrective or "pre-hab" exercises
4. movement preparation or active stretching
5. strength training
6. cardio or "metabolic conditioning"
7. stretching (passive)
8. nutrition

Yes, I put nutrition in there at the end because, at least for me, it is vitally important to feed my muscles nutritious food, including both carbs and protein, soon after exercise.

JULIE DESCH, MD

Experts say a ratio of 4:1 carbohydrate to protein is what you should shoot for. You can do this very easily by drinking some chocolate milk or eating a peanut butter sandwich. It doesn't have to be complicated, but it does have to happen.

So, let's go through the actual workout. Note: More description and even demonstrations of some exercises can be found on my blog at www.sickandhappy.com.

Soft Tissue:

I do about 10 minutes of foam rolling or use a tennis or lacrosse ball in areas of muscle and connective tissue that is excessively tight. You know an area needs to be rolled when it is uncomfortable to do so. If you don't feel discomfort, you are good to go on to the next spot. I generally start with a ball to my feet (ouch), and then go to the foam roller for calves, hamstrings and glutes. I then flip over and do my quadriceps. Then I take out the magic "peanut," my extremely complicated and expensive device that I now absolutely cannot live without (wrap two tennis balls together with duct tape). With this, all my tension dissipates from my back, as it remembers how to extend after my day of sitting, coughing, and typing. That's it!

As I said, I'm fifty. I have accumulated a lot of tight areas. You may not need this much, or you may need more. Only you will know, by trial and error.

Mobility Exercises:

This is also not complicated, and we are talking about joints here. Having good mobility simply means that you are able to take each joint

through its natural range of motion. Each joint is different, of course. The knee joint shouldn't be able to traverse a circle, while the ankle joint should (ha...tell that to my ankles!). Take a survey of your body. You will be able to tell which joints are tight. Work on those. Also, do some active range of motion drills in the joints that you intend to use in your workout.

Corrective Exercises:

These are also referred to as "prehab" exercises, presumably because if you do them, you won't ultimately require "rehab" exercises. Simply put, the idea is to strengthen weak areas that contribute to unhealthy movement patterns or poor posture. In my case, and likely in the case of anyone with CF, this is primarily my thoracic spine. Lung disease and chronic coughing cause the biomechanics of the chest wall to get messed up (to use a technical term). The result is the "hunched" back and rounded forward shoulders we commonly see in each other. Corrective work for this focuses on opening the anterior chest and shoulders with active stretching, and strengthening the muscles of the back that pull the shoulder blades back and down.

Movement Preparation (AKA active stretching)

This is the "warm up" part of the workout. The goal for this portion of time is to actively work the areas of your body that you are about to engage. You slowly start asking more of the heart and lungs as you begin using large muscle groups in a similar way to what you are about to ask of them. For example, if this is a leg workout day, you might begin with some lunges or body squats and add in some walking hamstring stretches. If you are going to focus on bench pressing, simply pressing a light weight for a few sets of 5-8 repetitions would be a great warm up. If you are going for a walk or jog, beginning to do that exercise at a slow rate for a few minutes is the way to go. It all depends on what you plan to focus on that day.

Strength Training or Cardio (metabolic conditioning)

I put these in the same category because I would suggest focusing on one or the other during a workout. You can do this by alternating lifting days with cardio days. Alternately, you can do both at the same time by doing weight training in a circuit fashion, with little rest between exercises. My favorite way of doing this these days is with kettlebells, which I will discuss in another article.

The main thing to remember here is to start small and slowly progress as your body adapts to the challenge. If you want to be able to run a 10K, that is awesome, and you can do it! But start with walking/jogging intervals which feel like work, but also feel good! There is no better way to sabotage yourself than to rush your body faster than it can go. How will you know if you are rushing things? You won't want to keep doing it. When you start dreading your daily jog, you know you are pushing too hard.

Stretching (Passive)

Ah...this is what you've been waiting for. The end of the workout! You've done your last set, or run your last interval. You want to grab your stuff and go fall on your couch. But wait! There's more...

This is the time to do just a little bit of flexibility training. Your muscles are warm and pliable...a perfect set-up for some passive stretching. Spend just 5 minutes stretching those areas on your body that tend to be tight. You don't need to go through a whole yoga series here. You know what you need. I almost always need low back and hamstring stretches at this point. I also find that this is the perfect time to lie across the foam roller lengthwise (so it is under my spine from my head to my butt) and open my arms to the side and let gravity open my chest. Some deep, meditative breathing in this position is the perfect way to conclude the workout.

And before you hit the couch, don't forget to eat! ▲

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

TRANSPLANT TALK

Surviving The Median Age

By Andrea Eisenman

I remember back when my mom used to sit down with me and explain why I had to do an inhalation. I must have asked her, "Why?" a lot, as I remember having this conversation with her many times. I could not understand why my friends didn't have to do inhalations, or take enzymes before they ate, or didn't cough all the time. I kept asking things like, "I won't have to take these pills when I get older, right?" Her answers, which were, "Yes, you will have to take pills when you get older, you have a mild form of a disease. It does not get better." didn't make so much sense to me.

At that point, I am sure I had no idea what "mild disease" I had. I just knew I had horrible stomach aches and hated taking enzymes and antibiotics. I suggested we dispense with giving them several times a day and that I take them all at once. It seemed reasonable to me, at six years of age. My mom told me, "No, it doesn't work that way." The pills didn't even make me feel better. They were what she called "preventative". I thought, you get a headache, you take an aspirin and the headache goes away. Why didn't my symptoms go away? I was a late bloomer and I don't think it was until I was about 11 or 12 that I wanted to know more about this "mild disease".

Little did I know, the more I would learn about cystic fibrosis (CF), the more I would wish I didn't know. Like, specifically, the median age of survival. I was one of those people born in the '60s, when pediatricians said to parents, "You will be lucky if your child lives until six-years-old." (This I found out later.)

I was angry that I had this disease and angry about how it made me different from my friends. People asked me all the time about whether I was sick with a cold, due to my cough. I told them I had a disease called cystic fibrosis - and no one had ever heard of it. They looked more confused than when they asked if I had a cold. I just started telling people I had asthma, to simplify things and to keep people from avoiding me.

> *"The more I would learn about cystic fibrosis (CF), the more I would wish I didn't know. Like, specifically, the median age of survival."*

But when I was 13 and in high school, in my biology class the teacher was talking about cystic fibrosis. I suddenly wanted to tell someone, "That is what I have, not asthma!" And then the teacher said, "It is a childhood disease. The age of survival for those who have CF is 13." I was a bit crushed and there started my nihilist phase of - nothing mattered, I was not going to make it to 18. Why bother studying or getting good grades? Why couldn't I smoke cigarettes or get stoned on marijuana? That was the beginning of my angry and rebellious phase.

I am embarrassed to report that I did smoke cigarettes for a year and pot for several, before I finally stopped. By then, the damage was done. The marijuana was like self-medicating to control my feelings of helplessness over having CF and not knowing others with the disease. I felt isolated and did stupid things. Whenever I read about CF or heard things on TV, I was always at the median age of survival. It was like growing up without a future. I felt college was a waste of time, as my time was limited.

Thankfully, I snapped out of that, due to my mom convincing me to go to college. I went for four years and received a BFA in art. I did go to a state school thinking that I was not worth the money to spend on a private education. Little did I know then, it is not so much what you learn from books or studying, but what you learn about yourself and the world. It helped me a great deal to go away to school and be on my own. I became more responsible for myself and doing my treatments.

My mom told me to stop reading things about CF in encyclopedias as they were outdated and; besides, I had a mild case. She tried to get me to understand, what the doctors tried for years to drill into my head: if you do your treatments, it will prolong your life. Don't go by statistics. But even the CF Foundation, who I looked to for answers, marketed CF as a "childhood disease". Later I would realize that this was probably to get more sympathy and donations for children who would die young, rather than snotty, rebellious teenagers dying young or trying to kill themselves by smoking.

I ask myself frequently if, knowing that CF was not a childhood killer, that my longevity was undetermined, would I have made the choices

I made? I would hope so and, also, hope not, since I cannot go back and relive my life by making smarter decisions. And I would never want to go through puberty or taking the SATs again! But I do feel like it nags at me.

I have had a very different perspective, since I was 23. The doctors told my parents that I would not live more than six months, take me home and let me do what I wanted. Even though I did not consciously know this information, I knew I had to change my ways. I then prioritized my health over a social life. Work became a priority, as I needed health benefits from my employer. I took care of myself to be able to work and worked to get insurance. I was eventually able to adjust and fit in a smaller social life. If only I had prioritized my health when I was younger, but I guess I felt it would not have made a difference--I had a "deadly disease". I am sure now that it would have made a difference in my health.

At 34, while on the waiting list for a bilateral lung transplant, I had finally stopped thinking about the median survival. Once while seeing my pulmonologist, during a routine check-up, I asked him about the chances of surviving the transplant operation. He stated that even if my transplant was not successful, I had outlived the median age of CF. That I should feel good about that. I found his answer to be a bit 'flip' but realized that I finally felt it was a more individualized disease than a death sentence. I might have taken it as a compliment because I knew he was impressed that I was still alive with how damaged my lungs were at that point. He must have felt I was doing something right. If he knew only the younger version of me, he would shudder.

Of course, once I had the transplant my life changed drastically for the better. I then focused on the survival rate for lung transplants being only, at that time, 85 percent after the first year and less than 50 percent for the next two years. I was, seriously, a nervous wreck until after the first three years. I was worried all the time about rejection and infection, and I was being extremely compliant. I realized I had to stop going by statistics (again!), live my life fully and enjoy it. I was given a second chance.

Thankfully, since my diagnosis in 1965, at nine months of age, the new therapies and scientific breakthroughs for CF have been staggering. Now, people born with CF today, and in recent years, have a much brighter future. With the median age over 38, I hope people will not fall into the same anger/pity spiral I did, but feel they can be the ones who outlive the median age when it is 80! ▲

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

WEBcasts
Sponsored by the CFF

Check in regularly at www.cff.org for information about Virtual CF Education Day Webcasts, sponsored by the CF Foundation. The most recent Webcast will feature CF-Related Diabetes (CFRD): What You Should Know. Check out the Website for more information: www.cff.org.

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.

ARNOLD *continued from page 1*

able to give you the proper care. Ask if community shelters can accommodate your needs and find out if you must register for them.

Include evacuation routes and safe places in your plan. Know where to go in each type of disaster. Know the location of fire extinguishers, disaster supply kits, and first aid kits. Know where the separate utility shut-offs are in your home and know how to safely and properly utilize them.

You should assemble disaster supply kits to include but not limited to:

Basics supplies

- Nonperishable food items
- A three day supply of water
- Flashlight and radio with additional batteries
- Extra clothing for cooler climates
- Sanitation and hygiene products
- First aid kits
- Matches

Medical supplies

- Medical alert bracelets
- A copy of your medical records and important patient health information
- A two week supply of your medications
- Medical devices and supplies

When selecting non perishable food products, you should stock a well balanced variety and remember to consider dietary restrictions. Many people who have CF have diabetes and should avoid stocking up on foods that are high in sugar.

Generators and other energy sources would be beneficial so that you may use your nebulizers in case of long-term power outages. Remember to take your medications and stay adherent because failure to do so could lead to exacerbations that may add more stress to an already stressful situation.

Design separate plans for different types of disasters; and remember to review your plans from time to time in order to remain familiar with them as well as keep them current. While we can't prevent disasters we can become proactive and take precautions to minimize their impacts.

Resources

- Preparing for Disaster for People with Disabilities and other Special Needs: http://www.redcross.org/images/pdfs/preparedness/A4497.pdf
- Disaster Preparedness for Persons with Chronic Disease: http://www.chronicdiseasepreparedness.org/pdf/patientresourcemanual.pdf ▲

Information from the Internet...

Compiled by Laura Tillman

This issue brings a potpourri of articles from the Internet

NEWS RELEASES

APP Pharmaceuticals Receives Approval for Piperacillin and Tazobactam for Injection

APP Pharmaceuticals, Inc. announced that it has received approval from the U.S. Food and Drug Administration to market Piperacillin and Tazobactam for Injection, the number one prescribed intravenous antibiotic in the U.S. APP will launch the product immediately.
http://tinyurl.com/3kgojyz

Battle of the Bacteria: Genetically Modified E.coli vs. P.aeruginosa

Biochemical engineers have genetically modified a strain of the Escherichia coli (E. coli) bacteria to fight Pseudomonas aeruginosa (Pa). By inserting DNA fragments into the E. coli, the engineered bacteria strain produced pyocin S5, a toxic protein which kills Pa strains. Pyocins are produced by Pa itself to compete with its own species, so they are species-specific antibiotics. Researchers also engineered their E.coli strain to only release the pyocin when it detects Pa bacteria. They exploited the organic chemical signaling cascade of Pa and engineered their E.coli in a way such that it would release pyocins only when it detected the chemical signals that Pseudomonas bacteria send to each other.
http://tinyurl.com/3bm3q4o

Lamellar Biomedical Secures Its First Orphan Drug License

Lamellar Biomedical Ltd announced that their lead clinical candidate, LMS-611 for the treatment of patients with CF, has received Orphan Drug Designation from the European Commission. Lamellasome therapies act on contact to disrupt the thick, sticky mucus which clogs the airways, making the clearing of mucus easier, reducing the need for physical therapy, and improving overall breathing function. Lamellasome therapies also disrupt bacterial colonies, overcome infectious biofilm buildup and improve the efficacy of certain antibiotics up to 16-fold.
http://tinyurl.com/3m6jwxh

Two Promising Projects Focus On New Treatments For CF

Two promising projects have been selected to undergo preclinical testing

Continued on page 28

CFLF Offers Grants For Recreation

The Cystic Fibrosis Lifestyle Foundation (CFLF) is a nonprofit organization that is based in Burlington, VT. The ongoing mission of CFLF is to provide Recreation Grants to assist CF patients, nationally, with activity costs as a form of physical, psychological and social therapy. Activities that have been approved for funding in the past include, but are not limited to, gym memberships, yoga classes, swimming lessons, martial arts, dance classes, gymnastics, golf fees and summer camps.

CFLF
Cystic Fibrosis Lifestyle Foundation

Brian Callanan, the Founder and Director of CFLF, is 35 and has CF. He lives in Vermont. He created the organization in 2003, with the strong belief that physical activity for people with CF is not only important but vital for improving and maintaining the overall health and attitude of individuals with CF. Assisting Brian is Program Coordinator, Erin Evans, 28, who also has CF. Both she and Brian are passionate about staying active through recreation and believe that living stronger *is* living longer.

CFLF currently is seeking individuals who are interested in golf, gymnastics, dance, horseback riding and/or swimming to apply for the Loretta Morris Memorial Fund. Born March 31, 1950, Loretta Morris died at age 21 in November 1971, while attending California State University at Northridge. She was not diagnosed with CF until she was 16. In spite of her illness she enjoyed the recreation activities of horseback riding and dance and also was an avid reader. In honor of Loretta, her sister, Barbara, has established this fund to help people share in activities both Loretta and she enjoyed.

Preference for this grant will be given to California residents who are interested in the above activities, but funding is NOT strictly limited to these areas. ALL applications will be seriously considered.

To learn more about CFLF and other grant opportunities and to download an application, please visit: www.CFLF.org or make direct contact at: Brian@CFLF.org or Erin@CFLF.org

Encourage Family and Friends to Sign Donor Cards

Give the gift of life that lives after you.

To receive donor cards, call:

United Network For Organ Sharing 1-800-355-7427

Isa: I would encourage my CF peers awaiting transplant to keep a list of all the things they want to do and see in their lives. Hope and goals have kept me going, and I would hope the same for my peers. I'd suggest a lot of prayer and inner work to prepare for potential tough times ahead, and to BELIEVE in one's strength and resilience. If you are depressed, that's normal! I'd suggest asking about happy pills, as there is no need to suffer more than necessary. I'd definitely encourage people to write their stories, which can help deal with the emotional struggles or trauma we naturally go through when faced with transplant. Or, I'd suggest at least scrapbooking their experiences, to document this phase of their journey. I'd highly recommend exercise, as that's what got me through the toughest phase of transplant. I'd definitely tell these peers to write to their donor families afterwards.

Andrea: As one of the interviewers who was fortunate to see the movie, I highly recommend *The Power of Two*, the documentary. I felt it truly captured having CF plus living with a life-changing bilateral lung transplant and, in Ana's instance, twice. I came away feeling that we can make changes in our lives or as a collective using our stories for advocacy. It is about educating people to take the fear out of the unknown and leading the way, which this film exemplifies beautifully.

We are all stars, whether we have CF, have a transplant, are a caregiver, family member, CF care provider, or friend. We are looking to shine our "spotlight" on other willing participants. Please email us to be interviewed.

Jeanie Hanley: jhanley@usacfa.org
Andrea Eisenman: aeisenman@usacfa.org ▲

TILLMAN *continued from page 26*

as part of the Cystic Fibrosis Technology Initiative. One project uses a new combination of antibiotics and chemical elements to target bacteria, while the other focuses on the cycle of inflammation and infection that damages the lungs of CF patients. The first project features a new formulation of an existing antibiotic, ciprofloxacin, combined with gallium to produce a potent therapeutic against infections. The second project focuses on a unique peptide from a family termed defensins. This short peptide holds the promise of being both anti-inflammatory and anti-bacterial.
http://tinyurl.com/3kbccxv

Gene Splicing Breakthrough Gives Hope To CF Sufferers

Researchers have discovered that an RNA molecule previously thought to play no role in gene splicing is essential. Researchers have also discovered a new way to study what the U4 molecule does. Gene splicing involves the cutting out of unnecessary portions of a gene. The remaining parts of that gene then join together. Although the process occurs naturally, scientists are able to mimic it. A new way to study how the U4 molecule acts inside a test tube has been developed. The system is called reconstitution. The idea is that you get rid of the U4 molecule that's normally there and replace it with one of your making. That allows you to test a variety of mutations. Parts of the molecule can be removed or changed around.
http://tinyurl.com/43to6f8

Pseudomonas Deploys A Toxin Delivery Machine To Breach Cell Walls Of Rivals Without Hurting Itself

Microbiologists have uncovered a sneaky trick by the bacterium *Pseudomonas aeruginosa* (Pa) to oust rivals. It deploys a toxin delivery machine to breach cell walls of competitors without hurting itself. Its means of attack helps it survive in the outside environment and may even help it cause infection. The scientists discovered that Pa injects toxins into rival bacteria with a needle-like puncturing device called the type VI secretion system (T6SS). The toxins degrade competitors' protective barricades – their cell walls. After the cell wall is compromised, the cell bursts like an overfilled water balloon. The T6SS mechanism transports toxins so that they never enter Pa's cell wall space. To thwart an attack from other members of its species, each Pa cell also has specific immunity proteins that inactivate toxins injected by neighboring cells.
http://tinyurl.com/3t3k3r4 and http://tinyurl.com/3m599h2

Pulmatrix Wins $14M to Back Drug

New Online Nomination Form!

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

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

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

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

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

For CF And Other Lung Ailments

Pulmatrix announced that it will advance its lead compound, called PUR118, as a new treatment for CF. PUR118 is a dry powder that patients can inhale. Pulmatrix is using its technology platform, which it calls iCalm, to create aerosol treatments that attack lung ailments from three directions. First, they have positively-charged ion-based compounds, like calcium and magnesium, which stimulate the immune system to fight off many different pathogens that people breathe in all the time, like viruses and bacteria. Secondly, the drugs produce an anti-inflammatory effect without the use of steroids. And finally, they help clear mucus from the lungs.
http://tinyurl.com/3t7ml6b

Ataluren Phase 2 Data In Nonsense Mutation CF

Ataluren (formerly referred to as PTC124®) is a protein restoration therapy designed to enable the formation of a full-length, functional CFTR protein in patients with genetic disorders caused by a nonsense mutation. A nonsense mutation is an alteration in the genetic code that prematurely halts the synthesis of an essential protein. The published three-month data showed that treatment with Ataluren resulted in statistically significant improvements in chloride channel activity, CF-related cough and positive trends in lung function. The results are important because they suggest that Ataluren promotes the production of a full-length, functional CF transmembrane conductance regulator (CFTR) protein and addresses the underlying cause of the disorder.
http://tinyurl.com/6echzlj

INFECTION CONTROL

Infection Prevention And Control In CF.

Saiman, Lisa. Current Opinion in Infectious Diseases. August 2011 - Volume 24 - Issue 4 - p 390–395

This review discusses recent studies of the virulence of CF pathogens, including epidemic strains, a more complex understanding of droplet transmission, bacterial contamination of CF clinics, and identifying and overcoming barriers to implementation of infection control guidelines. Recent experimental and clinical data have suggested that CF patients can generate droplet nuclei in the respirable range and that infectious particles can be cultured from room air minutes to hours after patients have left. Although these observations regarding droplet nuclei have not been linked to transmission, they do challenge the safety of the "3–foot rule" and suggest the potential role of masks.
http://tinyurl.com/3syjnsb

COMPLICATIONS

Gout And Hyperuricaemia In Adults With CF.

Alex Horsely, Jennifer Helm, Amanda Brennan, Rowland Bright-Thomas, Kevin Webb, Andrew Jones. Journal of the Royal Society of Medicine. 2011;104:S36-S39

Gout has not been described previously as a complication in CF. Data on nine CF patients who have presented with symptoms of acute gout are presented. This gives an estimated prevalence of gout of around 2.5% in the adult CF population, compared to a previously described prevalence in the non-CF population of just over 1%.
http://tinyurl.com/42tvb3y

Risk Factors for Chronic Kidney Disease in Adults with CF.

Bradley S Quon, Nicole Mayer-Hamblett, Moira L Aitken, Alan R Smyth, and Christopher H Goss. American Journal of Respiratory and Critical Care Medicine. Published ahead of print on July 28, 2011.

CF–related diabetes is a significant risk factor for chronic kidney disease in adults with CF, but additional studies examining IV aminoglycoside exposure directly are required.
http://tinyurl.com/3s2v8uf

Lumbar Disc Herniation In Three Patients With CF: A Case Series.

Christian Denne, Anna E Vogl-Voswinckel, Harald Wurmser, Marc Steinborn, Manfred Spaeth, Armin Gruebl and Stefan Burdock. Journal of Medical Case Reports. 2011, 5:440

Lumbar disc herniation can lead to a vicious cycle for patients with CF as it may promote pulmonary infections.
http://tinyurl.com/3nr34vg

TREATMENTS

Reduced Mortality In CF Patients Treated With Tobramycin Inhalation Solution.

Gregory S. Sawicki MD, MPH, James E. Signorovitch, Jie Zhang, Dominick Latremouille-Viau, Markus von Wartburg, Eric Q. Wu, Lizheng Shi. Pediatric Pulmonology. Article first published online: 3 AUG 2011

Though tobramycin inhalation solution has been used for over a decade to improve lung function and reduce exacerbations in patients with CF, its effects on mortality have not been well-described. After adjustment for multiple patient characteristics and known risk factors, use of tobramycin inhalation solution was associated with significantly reduced mortality among patients with CF.
http://tinyurl.com/3v3n4k9

Results Of A Phase IIa Study Of VX-809, An Investigational CFTR Corrector Compound, In Subjects With CF Homozygous For The F508del-CFTR Mutation.

J. P. Clancy, Steven M. Rowe, Frank J. Accurso, Moira L. Aitken, Raouf S. Amin, Melissa A. Ashlock, Manfred Ballmann, Michael P. Boyle, Inez Bronsveld, Preston W. Campbell, Kris DeBoeck, Scott H. Donaldson,

Continued on page 30

Henry L. Dorkin, Jordan M. Dunitz, Peter R. Durie, Manu Jain, Anissa Leonard, Karen S. McCoy, Richard B. Moss, Joseph M. Pilewski, Daniel B. Rosenbluth, Ronald C. Rubenstein, Michael S. Schechter, Martyn Botfield, Claudia L. Ordoñez, George T. Spencer-Green, Laurent Vernillet, Steve Wisseh, Karl Yen, Michael W. Kinston. Thorax**.** Published Online First 8 August 2011

Pharmacodynamic data suggested that VX-809 improved CFTR function in at least one organ (sweat gland). VX-809 reduced elevated sweat chloride values in a dose-dependent manner that was statistically significant in the 100 and 200 mg dose groups. There was no statistically significant improvement in CFTR function in the nasal epithelium as measured by nasal potential difference, nor were there statistically significant changes in lung function or patient-reported outcomes.

http://tinyurl.com/3uev5tj

Longitudinal Association Between Medication Adherence And Lung Health In People With CF.

Michelle N. Eakin, Andrew Bilderback, Michael P. Boyle, Peter J. Mogayzel, Kristin A. Rieker. Journal of Cystic Fibrosis. Volume 10, Issue 4, Pages 258-264, July 2011

The results demonstrate a significant relation between medication adherence and IV antibiotics in CF patients, highlighting the importance of addressing adherence during clinic visits to improve health outcomes.

http://tinyurl.com/43t32rq

Aztreonam For Inhalation Solution (AZLI) In Patients With CF, Mild Lung Impairment, And Pseudomonas aeruginosa.

C.E. Wainwright, A.L. Quittner, D.E. Geller, C. Nakamura, J.L. Wooldridge, R.L. Gibson, S. Lewis, A.B. Montgomery. Journal of Cystic Fibrosis**.** Volume 10, Issue 4, Pages 234-242, July 2011

Effects on respiratory symptoms were modest; however, forced expiratory volume in 1 second (FEV_1) improvements and bacterial density reductions support a possible role for aztreonam for inhalation solution (AZLI) in these relatively healthy patients.

http://tinyurl.com/5raj3r6

Inhaled Levofloxacin Beneficial In CF Patients With Pseudomonas aeruginosa Infection.

Neil M. Ampel, MD. Am J Respir Crit Care Med 2011 Jun 1; 183:1510

In a placebo-controlled trial, inhaled levofloxacin (240 mg twice daily) significantly reduced sputum *Pseudomonas aeruginosa* density and improved pulmonary function.

http://tinyurl.com/5w8dxfw

FYI

Why Infection With A Mycobacterium Is Increased By Long-Term Antibiotic Use.

Maurizio Renna, Catherine Schaffner, Karen Brown, Shaobin Shang, Marcela Henao Tamayo, Krisztina Hegyi, Neil J. Grimsey, David Cusens, Sarah Coulter, Jason Cooper, Anne R. Bowden, Sandra M. Newton, Beate Kampmann, Jennifer Helm, Andrew Jones, Charles S. Haworth, Randall J. Basaraba, Mary Ann DeGroote, Diane J. Ordway, David C. Rubinsztein, and R. Andres Floto. The Journal of Clinical Investigation. Volume 121. Number 9; September 2011

A recent study indicated that azithromycin treatment in patients with CF is associated with increased infection with nontuberculous mycobacteria. A team of researchers has confirmed that long-term use of azithromycin by adults with CF is associated with infection with nontuberculous mycobacteria and identified an underlying mechanism.

http://tinyurl.com/3zjjwga and http://tinyurl.com/42b6dqn

Pain Is A Common Problem Affecting Clinical Outcomes In Adults With CF.

Margaret Hayes, M.D., Myron Yaster, M.D., Jennifer A. Haythornthwaite, PhD, Kristin Riekert, PhD, Kristen Nelson, M.D., Elizabeth White, R.N. M.S.N., Peter Mogayzel, M.D. PhD and Noah Lechtzin**.** Chest**.** June 2011

Pain is common in adults with CF, interferes with activities and is associated with lower quality of life and an increased risk of both exacerbations and death.

http://tinyurl.com/3f5vepy

Exacerbation Frequency And Clinical Outcomes In Adult Patients With CF.

Kaïssa de Boer, Katherine L. Vandemheen, Elizabeth Tullis, Steve Doucette, Dean Fergusson, Andreas Freitag, Nigel Paterson, Mary Jackson, M. Diane Lougheed, Vijay Kumar, Shawn D. Aaron. Thorax. Published Online First 15 June 2011

Patients with CF with frequent exacerbations appear to experience an accelerated decline in lung function, and they have an increased 3 year risk of death or lung transplant.

http://tinyurl.com/3zh7r5k

Infection Of Polarized Airway Epithelial Cells By Normal & Small-Colony Variant Strains Of Staphylococcus aureus Is Increased In Cells With Abnormal CFTR Function & Is Influenced By NF-B.

Gabriel Mitchell, Gilles Grondin, Ginette Bilodeau, André M. Cantin and François Malian. Infection and Immunity. Published online ahead of print on 27 June 2011

The study supports the hypothesis that the pro-inflammatory status of CF tissues facilitates the infection of pulmonary epithelial cells by S. aureus.

http://tinyurl.com/3vxuo9x ▲

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

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