The Cystic Fibrosis Foundation Announces New Initiative For Adult Care

In March, the Cystic Fibrosis Foundation (CFF) announced the launch of a new $5 million initiative to enhance care for the growing population of adults with cystic fibrosis (CF). The initiative, called the Program for Adult Care Excellence (PACE), will expand the scope of adult care programs for people with CF.

The number of adults with CF continues to increase as life expectancy for the disease continues to rise. To meet the growing demand for care, the CFF will recruit and train CF care providers and fellows, with an overall goal of adding 40 new adult providers in care centers nationwide.

“The need for enhanced resources to treat the adult CF population is a direct result of our success in extending the life span of people with cystic fibrosis,” said Preston W. Campbell, III, M.D., executive vice president for medical affairs of the CFF. “What used to be a pediatric disease is now increasingly a chronic illness that brings its own set of challenges for adults.”

In 1955, when the CFF was established, children with CF rarely lived long enough to attend elementary school. Today, thanks to improvements in CF research and care, the median predicted age of survival is 37, and 43 percent of all people with CF are over the age of 18.

Over the past two decades the CFF has made key investments in adult care to keep pace with the growing adult population. For exam-
A WORD FROM THE PRESIDENT...

The Directors of USACFA, just as anyone else, sometimes have family issues to deal with. Some of us are caring for ailing parents or dealing with our own health issues. Some are dealing with both. We find that we must fit the needs of USACFA and CF Roundtable into the needs of our own lives. I know that you all understand that. Fortunately, we all are able and willing to fill in as needed. For that reason, I am writing this column at Paul Feld's request. His father has been very ill and Paul is busy with those obligations. We wish his dad a speedy return to better health and we look forward to reading Paul's writings again.

We are sad to report that Ken O'Brien, a founder of USACFA, died on March 2, 2008. See his obituary on page 25. Ken was the first Secretary of USACFA and an early Editor of CF Roundtable.

This issue has many articles to catch your interest. Julie Desch writes about nutrition, in Wellness. In Transplant Talk, Colleen Adamson tells of her experiences with skin cancers. Kurt Robinson talks of his love of basketball, in Voices from the Roundtable. In Spirit Medicine, Isabel Stenzel Byrne discusses love and CF. Beth Sufian discusses the Family Medical Leave Act and private disability benefits, in Ask the Attorney. Rich DeNagel interviewed a Nurse Coordinator for Unplugged. Laura Tillman reviewed Heather Summerhayes Cariou's book, “Sixtyfive Roses A Sister's Memoir”. Laura also compiled the Information from the Internet column.

The Focus topic of this issue is: Traveling with CF. Kris Dopher, Andrea Eisenman, Jeanie Hanley and Maggie Sheehan all discuss various aspects of travel. In A Deep Breath In, Debbie Ajini writes of the limits that CF can place on one and on one's travels. Kathy Russell writes of another take on travel, in Speeding Past 50.

Be sure to check out the announcements and the article about Heroes of Hope. We hope you will nominate someone as a hero.

Please check out the Looking Ahead information on page 5. All of the Focus topics for the coming year are listed there. As always, we invite you to write for the newsletter. Anything that is CF-related can be considered for publishing.

We hope that you and yours are well and that all of us and ours soon will be too.

Stay healthy and happy,
Kathy Russell
Information from the Internet...

Compiled By Laura Tillman

This issue brings a potpourri of articles from the Internet

FYI


In patients with cystic fibrosis, the cumulative risk of radiation-induced cancer resulting from routine annual lung computed tomography (CT) is “quite small” — less than 0.5%, a study shows. However, the lack of documented benefit of routine CT makes its use questionable. Assuming current median survival of CF patients to age 36 years, the estimated risk of radiation-induced cancer from annual lung CT is 0.02% for males and 0.07% for females. If the median survival of CF patients improves to about age 50 years, as has been suggested, the estimated risk will increase to 0.8% for males and 0.46% for females. The risks are higher for females because of the risk of radiation-induced breast cancer (50% of total risk) and higher risk of thyroid cancer. The researchers conclude that routine CT monitoring should not be recommended until there is a demonstrated benefit that will outweigh these risks.


Gatifloxacin Produces Both Hypoglycemia and Hyperglycemia: A Retrospective Study. Haerian, Haleh MD; McHugh, Peter MD; Brown, Richard PharmD; Somes, Grant PhD; Solomon, Solomon S. MD. American Journal of the Medical Sciences. 335(2):95-98, February 2008.

Gatifloxacin, until recently one of the most commonly prescribed antibotics, has been shown to produce hypoglycemia. This study indicates that Gatifloxacin was clearly associated with both hypoglycemia and hyperglycemia. The risk of hyperglycemia increased in the presence of diabetes mellitus, steroid use, and “sick enough” to be in the intensive care unit.

http://www.mdlinx.com/PharmacistLinx/news/article.cfm/2141410/?user_id=188267&email=llt75@earthlink.net&news_id=658&subspec_id=267

NEWS RELEASE

Copernicus Receives Milestone Payment from Cystic Fibrosis Foundation Therapeutics to Further

Continued on page 13
1. I am in the hospital. Can my employer fire me while I am out sick?

The only real job protection while a person is sick, under Federal law, is provided by the Family Medical Leave Act (FMLA). A person must work one year at a job in order to be eligible for leave time under the FMLA. In addition, the person must have worked 1250 hours in the prior year and the employer must have 50 employees at offices within a 75-mile radius. If the employer is a bank and it has five branch offices within 75 miles of each other and employs a total of 50 people, then the FMLA will apply.

FMLA allows 12 weeks of unpaid leave to care for oneself, spouse, child, or a parent. If someone other than the person who is sick requests FMLA leave, the physician treating the person who is sick must complete the FMLA paperwork. A violation of the FMLA can be reported to the Department of Labor. A person’s employment should not be terminated while out on FMLA. However once the FMLA time has run out, the employer can terminate the employment if the person is unable to return to work at that time and does not have any other available sick or vacation time left. Employers can count sick and vacation time as part of the FMLA leave 12 weeks. For example, an employer could give two weeks of sick and vacation time and then 10 weeks of FMLA leave.

The employer can ask the employee to have a form completed by his treating physician indicating that the employee is out due to a serious medical condition. The FMLA defines a serious medical condition as one that requires either hospitalization or the care of a physician. If an individual does not want to disclose their CF diagnosis to their employer, the person with CF will have to discuss whether their physician feels comfortable writing a more general description of the person’s condition on the FMLA form.

The FMLA only protects an employee from having employment terminated because he is out sick or is out to care for a spouse, child or parent who is sick. The employer can terminate a person’s employment if the employee has violated company policy or broken the law, even if the person is on FMLA leave. For example, if an employee with CF is out on FMLA leave and during that time the employer finds that the employee had stolen money from the employer, the employer can terminate the employment even though the employee is on FMLA. The reason for the termination is not due to the election to take FMLA time but because the person broke the law.

Some people with CF mistakenly believe that the Americans with Disabilities Act (ADA) prevents an employer from terminating the employment of a person who is out sick. THIS IS NOT TRUE. Under the Americans with Disabilities Act most courts have held that a person who has used up sick and vacation time (and has either used up FMLA time or is not eligible for FMLA time) can be terminated from their job, if they are unable to work, even if the reason they are unable to work is due to an underlying medical condition.

Sometimes an individual may ask for reasonable accommodations under the ADA and request additional time off due to sickness. The courts have typically held that the employer does not need to give additional time off as a reasonable accommodation.
However, a person can certainly ask their employer if he will grant additional sick time as a reasonable accommodation. The chances of having additional time off provided increase, if other employees have been given additional time off.

2. How can I obtain short-term or long-term disability benefits from a private company?

Typically people with CF are not able to purchase short-term or long-term disability policies on their own. This is because there are no federal laws that require insurance companies to sell such policies to anyone who is interested. However, many large employers offer short-term and long-term disability polices to their employees. Often the employee must pay a small amount each month in order to obtain such coverage in the event the person becomes unable to work.

Typically, policies may require that the employee work for the company for a year before the policy will provide benefits, if the employee is out of work due to a condition that is pre-existing. People with CF should almost always opt for such a policy. Often the chance to enroll in coverage is provided only during the first 30 days of employment. After that time period the employee cannot enroll in the short-term or long-term disability plans. Employers have no duty to offer short-term or long-term disability to employees.

A person who is unable to work due to disability often is eligible for short-term disability benefits and long-term disability benefits, if the disability lasts a certain period of time. The provision of such benefits allows some salary replacement; typically it is 60-70% of salary. The amount of benefits will depend on the policy. However, being on short-term disability or long-term disability does not protect the person from having employment terminated. Usually, as long as the employee is on short-term disability before their employment is terminated, the employee will be able to receive short-term disability; and then long-term disability, if they are still unable to work – even if the employer has terminated their employment.

Beth is 41 and has CF. She is a Director of USACFA. Her contact information is on page 2. You may send her questions of a legal nature that are CF-related.
For the last 25 years, I’ve been involved with people in the cystic fibrosis community, and I would like to generalize a finding: we are an unusual bunch. I’m talking about all of us in the CF community; people living with CF, parents, partners and health care providers. While I cherish the individual differences each friend has to offer, overall, I’ve encountered the highest quality people.

I’ve heard a lot about modifier genes altering the expression of the CF gene, and hence the outcome of the CF prognosis. In my mythical thinking, many people with CF (and carriers) have modifier genes that produce proteins that attract kindness; that block channels of animosity; that overproduce the amino acids for love. These traits are all survival advantages.

I believe our day-to-day effort to stay alive creates a desperate bond, which makes us love deeper. I share this assertion of love in the CF community with a disclaimer. To be honest, I don’t belong to any other community the way I belong to this one. My hope is that the CF community reflects all other communities, like church or ethnic groups, merely mirroring the goodness in all people. Still, though, I think there is something special going on here.

We’ve been tagged with the branding iron of CF: a physical disease that wreaks havoc on our emotional, social, spiritual, romantic and sexual lives. Forced early by circumstance, we are challenged to adapt, to change, to find ways to be the best people we can be. I feel so blessed to witness a powerful love for life, self, and others in the CF community.

First, I believe our love affair with life is deeper compared to the average person. Our keen awareness of limited time causes us to cherish each moment, as clichéd as that sounds. I have marveled at crashing waves, a friend’s smile, petals on a flower, and I know many others with CF who share this deep attention to the world around them. My friends with CF have deeply inspired me because they’ve grasped life’s cornucopia of dreams with passionate love. When they travel to exotic places, pursue careers, have babies or transplants, I gain a firm conviction that if they can, I can.

For some, our love for life transcends our broken bodies and encompasses unseen powers. I have witnessed such strong spiritual auras from my CF friends, that when I’m in their presence, I feel like a privileged visitor from another place. This spiritual education has led me to believe that the love of the CF community reflects just an ounce of a greater love, God’s love. With my CF friends, the boundary between heaven and earth is blurred for me; but I still imagine a greater love awaiting us after this life. The love of something greater in life can lead to a sacred gratitude for the gift of being alive, no matter how short or compromised.

Our respect for life inspires healthy people around us. One mother who lost her son to CF was so depressed she felt no reason to keep living. But she remembered how her son and his friends with CF cherished each day and made the most of what they were given. She decided not to end her life and survived her grief. Later, she wrote me, “I thank the CF community for MY life. I thank the CF community for saving many people’s lives. You have given so many people a new perspective from which to live.”

Besides loving life, the hardest, but most important, love a person with CF can have is love towards oneself. Like many other people with CF, I’m often told that I’m courageous for living with a life-threatening disease. But, I think the true courage lies in peeling back the layers to unveil our...
authentic selves. Ovid said, “Fortune and love befriend the bold.” It takes courage to become real to self, then to others. Next comes the courage to love the good, the bad and the ugly parts of oneself; to love ourselves unconditionally. Then the gifts flow.

Personally, for many years, I experienced self-loathing because of my CF. This caused me as much suffering as my lung disease, until I realized it was self-inflicted. One day, I listed the positive aspects of myself, and I realized that many of them were developed because of my CF. This started the transformation of my negativity into self-love and acceptance. I am now on a decades-long pilgrimage trying to accept my body, abilities and life in general. This process of soul searching has been a long, hard path of forgiving myself for not being who I thought I was supposed to be. Many things have helped me love myself: a loving husband, working hard to stay healthy and having that pay off for three decades, a spiritual epiphany when my lungs failed, and a resurrection following lung transplantation. Most of all, writing about my CF journey and making sense of what happened has been extremely liberating. I’ve come a long way, but I still am learning to love myself. My favorite quote from Rumi says: “There is a way of breathing. That is a shame and a suffocation. There is another way. A love breath. That lets you open infinitely.” It is normal to experience anger towards oneself. But I have known a few CF friends who remain stuck there and project that anger to others, which only adds to the poison in their lungs. Even diseased CF lungs can take a love breath towards oneself.

While it is a lifelong challenge to love oneself, CF or not, I’ve seen people in the CF community possess an unusual love for others. Maybe this is because it is therapeutic to get outside of oneself and focus on others. I believe my disease molded how I relate to people. Whether it was how nurses treated me in the hospital or bullies at school, I learned that “honey attracts more bees than vinegar.” With a bit-tersweet belief in impending mortality, I tried to embrace and love those around me with sincerity. But usually, my treatment of others was just a reflection of how I was treated. I have been the recipient of tremendous kindness from my CF community friends, including CF parents. Whether it is a casserole at my doorstep, a phone call in the ER or a last minute dog-sitting when I was admitted to the hospital, I could not survive without the love of my CF community. Sometimes, I physically felt the love so strongly in my chest – a good kind of ache for a change – that I thought I’d explode.

I’m also touched by the love for others I see within CF families. CF forces some couples to sacrifice their own happiness and stay together to share in the care of their kids with CF. Some families are forced to go outside of their comfort zone and really communicate love. Partners love their CF partners for who they are. Parents who have CF endure the tough fight to see their children grow up. Bereaved parents and partners show us that their love can evolve and expand spiritually even after their CF loved ones have died. While this disease can break families apart, it can also create opportunities for families to love harder and deeper. This is what happened to mine.

One of my strongest bonds of love has been among fellow friends with CF. I’ve always had a desire to connect with people who’ve “been there.” I’ve seen love shine brighter when it is intertwined with the difficulties associated with CF: loneliness, insecurity, pain, fear and loss. When we drudge through this emotional mud we will sink unless we find solid ground to walk on. For me, that solid ground is my connection with - and my love for - others, which keeps me going.

Many of my CF peers are survivors and, hence, conveyors of hope. However, there were some friends whom I thought I might outlive, and the pain of knowing I would lose them made me love these friends in a more desperate way. CS Lewis said, “We love in order to know that we are not alone…to love at all is to be vulnerable. Love anything, and your heart will certainly be wrung and possibly broken.” Knowing that loss is inevitable, I’ve seen a pattern among CF friends: we don’t have time for grudges; we often forgive more easily. This has been so true for me.

You are likely reading this article because you or someone you love has a mutation on the seventh chromosome, maybe even F508 like me. Despite this genetic mistake, we got something right. There is nothing defective about our ability to love. In fact, the metaphorical modifier gene for love is practically heightened because of the CF defect. We have a choice of loving. How we love and whom we love defines our quality of lives, no matter how many years we have on this planet.

I hope this Spirit Medicine article encourages you to reflect on the love in your life and how CF has influenced that love. I recognize there are many people with CF who are yearning for love of life, self and others. I invite those who are lonely to take risks and become involved in the CF community. Come learn to give and receive. See what we have to offer… a rich community of love.

Inspired by NM and OD, my CF community friends.

Isabel Stenzel Byrnes is 36 and has CF. She is a co-author of “The Power of Two: A Twin Triumph over Cystic Fibrosis”. She lives in Redwood City, CA. She invites spiritual writers to share their ‘spirit medicine’.
When traveling as a kid, I used to see those giant Consolidated Freightways trucks with the huge “CF” on their side, and think they were cystic fibrosis trucks delivering massive amounts of food for children like me. I ate enormous quantities back then, so it didn’t seem like such a reach. Anyway, Consolidated Freightways long ago declared bankruptcy and is no longer in business, but this seemed like a great “travel” segue to the topic for today, eating.

As a reminder, in the past several issues, I have been writing about the concept of wellness by describing the body as an “energy transformer.” As humans, this is what we do. We take energy in and transform it into movement, thinking, working, playing, relating to others, etc.

The first article in the series dealt with the overriding principles of self-responsibility and love, which create a context within which the transformations occur. Then, we looked at the first two of three “energy inputs”, namely, breathing and sensing. Our third and final energy input is “eating.” As you know, the food you eat is digested and assimilated into the body in order to combine with the oxygen you breathe to form the energy required to provide heat and electrochemical energy. This, in turn, is used in the daily activities of life, as well as building and repairing tissues.

I have written about the nuts and bolts of nutrition in a previous article entitled Eat, Drink and Be Well, so I don’t really want this current article to be a discussion of macronutrients. Instead, I would like to look at eating from an energetic point of view. It may sound very “New Age,” but from an “energetic system” point of view, what we put into our body as “input” is very important.

Have you heard the adage “garbage in, garbage out?” Perhaps you have even experienced this concept on a level that only a malabsorption sufferer can?

If it is true that “garbage in = garbage out”, then it is also likely true that “healthy in = healthy out” (or something like that).

The last time you filled your car’s gas tank, did you pour in Coke, Kool-Aid, or nachos? No. It was likely nice clean, healthy $5/gallon gasoline that you trusted would keep your engine happy and running smoothly, for as long as you could afford to fuel it.

If you regarded what you put in your mouth as “fuel” for your body, and not just what sounds good to eat in the moment, would you eat differently? I know that when I remember to think this way, I eat differently. And when I eat consciously, I definitely feel better. And when I feel better, the food tastes better, and eating remains one of the great pleasures of my life.

This concept takes on even greater importance when we are sick. It is an evil irony of nature that we tend to lose our appetites and NOT want to eat anything when we are fighting a lung infection, breathing hard, and in need of even more calories than usual.

This winter, I experienced that feeling of complete absence of appetite to an extent that I was truly afraid I might never want to eat again. I was sick, in the hospital, and completely surprised every time the very nice nutrition services person would cheerfully take away my untouched tray of food, only to replace it with the next, completely unappetizing meal. Now, granted, this was hospital food…but even the
cookies my kids brought in, or my favorite Jamba Juice my friends brought in did not entice me. It wasn’t just disinterest. The thought of eating was rather nauseating.

At home, five days and eight lost pounds later, I knew that I couldn’t listen to my body anymore. I was wilting away and had to take a new approach. My very good friend had made me a huge batch of chicken soup as a welcome home present. If I could eat anything, it would be warm and salty. So I determined I would look at my “feedings” as I did my IV infusions. I wouldn’t have dreamed of skipping a dose of medication, feeling as horrible as I did. Likewise, I sat in front of some soup, and made myself at least taste it.

No, I didn’t immediately recover and start eating with my usual reckless abandon. In fact, I just made myself sit and at least taste food several times a day for a few days. Each time, I would remind myself that if I didn’t give my body any energy source, I would not get better.

Then, after about three days at home, I suddenly HAD to have a steak. The old Julie was back!

Why this long story? It’s simple really. The body lies. It tells you that you aren’t “hungry”…or to eat junk…or that ketchup counts as a vegetable. Sometimes, you just need to overrule and remind the body who is boss. You are in charge of what goes in, and You know best what it needs to function optimally.

Personally, I don’t think it works well for me to gauge water intake by thirst, or by some prescribed formula. I check out the color of my urine. If it is a pale yellow, I know I’m good to go. If it is too dark, I know I need to drink more. Similarly, gauge your nutrition status by how you feel. Do you have energy? Do you bonk in the middle of the afternoon? Are you starving an hour after your bowl of fruit loops?

Do some detective work. If you are like me, and most other people, you probably function best when you eat every three hours or so, and have balanced snacks including a source of protein, a healthy, whole food based carbohydrate, and some healthy fat (and your enzymes, of course). I bet if you try to eat a “rainbow” of colors every day, you will feel better. Don’t believe me, though. Try it out for yourself, and see what happens. ▲

Julie is 47 and is a physician who has CF. You may direct your questions regarding CF-related health issues to her at: jdesch@usacfa.org.

CFF and Vertex Announce Positive Early Results for VX-770

The Cystic Fibrosis Foundation (CFF) and Vertex Pharmaceuticals announced today (March 27, 2008) that VX-770, an oral drug in development that targets a basic defect in CF, showed promising results in an ongoing Phase 2a clinical trial for patients who carry the G551D mutation of CF. The drug is being developed by Vertex Pharmaceuticals Incorporated.

Patients who took the drug for 14 days showed significant improvements in several key indicators of cystic fibrosis, including lung function, nasal potential difference measurements and sweat chloride levels. The findings suggest that VX-770 improves function of what is known as the faulty CFTR protein. This early data is promising and could have important implications for studies of other drugs in development.

This is the first time that any potential therapy has improved the abnormal sweat chloride (salt) levels in a person with CF. Excessive sweat chloride is a key clinical indicator of cystic fibrosis. The “sweat test” is the traditional diagnostic test for CF.

“These early results are an extraordinary endorsement of our hypothesis—that small molecules can correct the basic defect and affect the clinical indicators of cystic fibrosis,” said Robert J. Beall, Ph.D., president and CEO of the CFF. “The emerging data for VX-770 represents the most exciting results we’ve seen from a Phase 2 trial and increase our confidence that we’re on the right track.”
I am delighted to realize that spring is upon us. I have survived another winter, which is not always an easy thing to do. Not only have I survived the winter, I have successfully completed another trip around the sun. That makes 64 times that I have made that orbit. When I was diagnosed with CF, in 1956, I know that no one would have believed that I would still be alive. But, here I am.

With our Focus topic being “Traveling With CF”, I find that the travel that means the most to me is the travel of time. Every year, hour, day, and second take on a special meaning when you have CF. Never can you just take for granted that life will continue and that all will be well. Each of us must make an investment in our own health. With any kind of good luck at all, we get to complete another trip around the sun and get to mark it with the addition of another year to our age.

For many people, getting older is nothing to celebrate. They fail to see the beauty of managing to survive. They take their trips in time for granted. They most likely never stop to think about how fortunate they are to age. They grumble about the signs of aging. They spend loads of money on preparations that are “guaranteed” to make them look younger. Not me!

I revel in my age. I am so happy to have the chance to see another day that sometimes I have trouble going to sleep, because I am so eager to see the next day. Each day brings new challenges and new victories and – sometimes - it brings new defeats. All of this is part of the “trip”.

When most people travel, they collect some kinds of souvenirs. The souvenirs from my trips around the sun are the wrinkles and graying hair that I sport. The aches in my bones and the surgical scars also serve to remind me of my “trips”. Each line and mark is my sort of tourist decal. (For those who are too young to remember “tourist decals”, they were regular transfer decals that were for sale in touristy spots and showed pictures of the places of interest. People used to stick them on their luggage or in their car windows. They were a way for people to say, “I’ve been here!”)

Those of us that have CF have our own special types of tourist decals. We sport many scars on our arms and hands, from the many IVs we have had. We may have scars from various kinds of surgeries, such as intestinal blockages, feeding tubes, problems with various organs, and more, including transplants. Each scar is a reminder of a part of our journey through life.

For members of the group of older people with CF, there are the memories of equipment that we used to use. Who can forget the mist tents, glass nebulizers, big compressors, smelly Mucomyst making everything sticky, sweat tests done with a large piece of plastic taped to one’s back overnight, duodenal drainages, old sinus surgeries done through the mouth, and the awful taste of old enzymes? These are more of our travel souvenirs.

Now life with CF is a little easier than it used to be. Compressors now are much smaller, quieter and easier to work with. Also, they often are battery operated or cordless and rechargeable. Our nebulizers are easily cleaned and are disposable. There are vest-type devices for chest physical therapy and they are portable. There are several different small devices for airway clearance. There are portable oxygen concentrators and small oxygen saturation analyzers. All of these things can make travel (in the conventional sense) a lot easier for those of us that are traveling through a CF life.

I doubt that many of us would say that this journey is easy or without its pitfalls, but most of us would say that we have learned from it. Because of CF, and my journey with it, I have met some wonderful people. My life would...
have been much less brilliant without those people in it. From them I have learned empathy, compassion, understanding, patience, tolerance, forbearance, and equanimity in the face of all manner of trials. Many of my friends have faced such great odds and have never let on how formidable those odds were. From them, I have learned to keep on keeping on.

I have not collected any replacement parts along my journey. I have lost a few of my original parts, but nothing I can’t live without. Each of those losses has created an improvement in my health status, so I am happy to have lost them. The scars from those losses are among my travel decals. I must say that I wouldn’t be sorry to have fewer of those travel decals. Each surgery means another round of anesthesia and we all know that we don’t want to be “put to sleep” any more times than we absolutely must be. (I firmly believe that we lose a few brain cells with each general anesthesia. I have no empirical evidence to back this up. It is just my opinion. I do know that I have lost memories of certain things, after surgery.) I guess that losing memories is another “travel souvenir”.

When I was early in this journey, I could get together with my friends who have CF and enjoy the fellowship and camaraderie of being with others who could finish my sentences about CF. We could laugh, cough, sputter and be ourselves, without worrying that we might “offend” someone. Everyone was in the same “boat”, so there was no concern about anyone not understanding. We often said that we should go en masse to a comedy club. Since most of us tried to laugh silently, to avoid causing ourselves to cough, the poor comics would think they were “bombing out”, when it would be just our odd way of laughing. We never did that. That would have been a great travel souvenir.

Now, we must keep a specified distance from each other. We tend to gather in places where we can meet outdoors or where we can have at least a chair between us. When we sit or stand next to each other, for the duration of a photograph, someone points out that we shouldn’t be so close to each other. We must keep our “three-foot-rule” all the time. Many who don’t have CF are very quick to point out our errors. I guess that is another of our travel souvenirs.

Along the way, I have had many friends die. Also, I have made many new friends. I cherish each friend, living or dead, and I am so happy that I could make the acquaintance of each of them. The photos I have of each of my friends are definitely among my travel souvenirs.

One of my most important souvenirs of my journey through this life is my husband, Paul. Paul has made my journey much smoother and definitely much more bearable. He is my advocate, my protector, and my best friend. He helps me keep my perspective and feeds my sense of humor. Without him, my life (with or without CF) would have been much poorer.

Am I through with my journey of CF? No way! Have I had the most interesting part of that journey? I have no idea. Do I want to know what lies ahead? Absolutely not! Do I look forward to the rest of the journey? Absolutely! Whatever happens and wherever this journey takes me, it is a wonderful journey and one that I am privileged to make.

I look forward to continuing my journey along with each of you. May our trips around the sun continue to be interesting—and it couldn’t hurt if they were filled with some good health. Please stay healthy and happy.

Kathy, 64, has CF. She is a Director of USACFCA and is the Treasurer. Her contact information is on page 2.

VOLUNTEERS NEEDED
FOR STUDIES AT NIH

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

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

We are looking for individuals with cystic fibrosis who previously participated in NIH studies. If you have taken part in an NIH study, please call the toll free number: 1-877-644-5864 and select #3 on the menu; or send an E-mail to: barnesp@nih.gov.
Boy, how times have changed for intrepid international travelers with CF! Even though my CF gets more challenging as I get older, the equipment improves so it really does seem to balance out in favor of easier travel overall. I’d like to give you my most interesting travel experience 10 years ago and compare it to today’s experience and what I’ve learned in between.

About 10 years ago, I traveled to France to meet my husband, who was on business there. The vests were larger then and not carry-on items, so I had to buy another shipping box in which to pack it. (The old shipping box had been used for a school project for my kids, before I realized that I needed it for traveling. Oops! But it was great for creating a puppet show.)

Also, at that time, my vest did not have dual voltage to automatically change to the European voltage like they do now. I had to buy a very expensive transformer/ converter to be able to use The Vest®. The day before I left, I realized that the converter was the wrong kind, because it converted from European to US instead of vice-versa. Needless to say, many dollars later, and a harrowing experience of having it delivered to the airport just before I was walking on to the plane, I had my transformer in hand.

I finally arrived safely in France, but after the plane ride, my nose was severely congested, my throat was too dry and my respiratory tract felt like someone had sucked out all the hydration. But who cared! I was in France! Ooh-la-la.

After several gorgeous days in Nice, my respiratory tract seemed to be responding well to the Mediterranean air. Around this time, my portable nebulizer/compressor had to be recharged. My unit had dual voltage capability, but you had to manually switch it over when it was being charged. (My new portable nebulizer automatically switches over.) No problem. I attached the French outlet adaptor, flipped the switch and charged.

Hours later, when the machine seemed deader than a doornail, I realized that I had already switched it over while on the plane and now I had completely, irreversibly killed the internal battery. Brilliant. We still had four days to go, but all ended well as a “pharmacie” nearby rented a prehistoric machine to me, (that sounded like a bulldozer, but worked) for an “asthmaticque”.

Fast forward to today with the automatic dual voltage vest and portable nebulizer. The Vest is lighter, more durable (I’ve seen baggage handlers merclessly toss it into the plane’s belly and, somehow, it continues to work well.) and can be carried on, if so desired. Lesson: all I have to buy for international travel is the appropriate
electrical socket adapter. The adapters must be bought in the U.S, as they are difficult to find outside the country.

I generally check-in the vest, except if I have a layover in Chicago O’Hare; then I will carry it on. The last time I flew through Chicago, all flights were cancelled due to poor weather. My vest had been checked-in and, although they tried to find every reason not to locate it for me, they gave in at the end (possibly a little coercion was used by my husband and me). All luggage had been placed in a holding area, not baggage claim, so it took them two hours to find it, which I thought was a small miracle because there were hundreds, if not more, pieces of luggage strewn about in these areas. A good description was necessary for them to find it. Lesson: due to frequently cancelled flights from poor weather at some airports, you’ll want to have all your equipment with you just in case you have to stay overnight waiting for another flight and better weather. Always place distinguishing markers on each piece of checked baggage, e.g. thick neon pink ribbon or brightly colored duct tape on all sides so that they can be found easily in baggage claim or elsewhere.

Since international travel is generally very time-consuming, the constant exposure to recirculated, dry air throughout the plane especially affects me. It causes extreme dryness in my nose, sinuses and lungs. There are several solutions that work well: I drink lots of water (I buy at least two 2-liter bottles at the airport for me and anyone else who’s traveling with me); cut off the airflow above my seat (and those around me if possible – I try to preboard for this reason) so that the air doesn’t blow powerfully up my nose. Also, I wear an iPod-sized humidifier like a necklace around my neck. I bought it at Sharper Image. It requires batteries, is very lightweight, easy to clean and, on a 12 hour flight to Italy, I needed to add only a small amount of distilled water once to the holding chamber inside. If you haven’t tried it and don’t mind a few strange looks from other passengers, then this is for you!

As for the dryness at the hotels, I have a similar routine (minus the humidifier) to prevent dehydration - drinking lots of liquids (not much alcohol since this dehydrates me), turning off the fan within the hotel room and opening windows for fresh air, if weather permits. Most importantly, I use a nasal saline lavage kit that can be bought or ordered from most pharmacies.

Shooting salt water up my nose did not used to be one of my favorite vacation activities. It took a while to get used to, but now I use it daily even when I’m not on vacation. I can’t seem to go without it, as its major benefits are fewer colds and sinus infections. This simple, convenient kit is small and, therefore, a great adjunct for travel. I use it at least two times everyday as it re-hydrates my nose and sinuses, reduces the number of germs/dirt and pathogens I’ve been exposed to during travel or visits to tourist areas, and overall keeps me well. Since it comes with premixed salt packets for use with the included bottle, it’s fairly user friendly.

International travel has definitely become easier. Our lifelines - our CF machines - thankfully have improved tremendously and thus improved traveling experiences. We still may have to visit a “pharmacie” or “farmacia” as it’s difficult to think of everything. So my last lesson would be this: if traveling to a foreign-language country, make sure you can pronounce key words (or have handy the phone contact of someone fluent) so that you can explain what you need. Bon Voyage! ▲

Jeanie is 45 and is a physician who has CF. She lives in Los Angeles with her husband, John, and their three teenagers.
By Cris Dopher

As Far North As I Can Go...
There’s traveling with CF, and then there’s TRAVELING with CF!

My blog entry of July 13th, 2007:

“A few days ago, I ran in Whitehorse, Yukon, where Robert Service wrote a couple of his favorite poems, including ‘The Shooting of Dangerous Dan McGrew’. Took some pics of the old surviving paddlewheel steamer they’ve preserved there. A short run; but much better than the one up at Muncho Lake. But this morning I ran at the Arctic Circle! It is not nearly as cold or barren as one would suppose. I know it’s high summer, but still...it’s warmer at the circle right now than in many places in lower Canada. I woke up to 70 degrees for my run, which turned out to be a hell of a hill workout. The elevation must be pretty high there. Anyway, it’s an interesting enough place; the Dalton Highway parallels the Alaskan pipeline and that is fascinating to look at - quite an oddity. And the whole tundra area is bizarre, especially now, since there was a 7 million acre wildfire recently and the hills are COVERED in fire-flower - a BRIGHT pink flower. So the whole area is a patchwork of green tundra grasses & trees and this pink wildflower. Great stuff! Haven’t dealt with the famous Alaskan mosquitoes, except at the arctic circle - they are minimal everywhere else, so far.”

Though I’m often pessimistic, my blessings are too numerous to count, truly. Key among them are these three: that I have cystic fibrosis, that I have a career which offers many opportunities to travel, and that I own a motorcycle.

Now, you may be wondering how I can count this disease as a blessing, but I do. I could go on at length about the positive aspects of CF, the benefits it accrues; but that’s for another time and another article. One element, though, is that CF propels me. With a constant sense of how short my time on earth may be, I feel I must make the most of the time I have. (This is nothing new to most CFers, I guess.)

What I choose to do with the time I have is to work in a rewarding profession and travel. Lots of travel.

I’ve purposely chosen a career that is the antithesis of the nine-to-five job. I am a freelance designer, with dovetailing professions as a CAD software teacher, draftsman, and writer. While a lot of this work can be done from home, it also involves frequent travel. For instance, I’m writing this on the train back from Blairstown, NJ, where a show featuring my set design is about to open. I’ve been back and forth for the last few weeks, painting the set for three or four days, then going back to New York for a few days. Round and round we go. And I’ve recently confirmed my teaching schedule, which has taken me to Vegas, Chicago, Bermuda, Calgary, Toronto, Vancouver, Seattle, and other interest-
ing locales. Point is: I’m getting pretty good at the five-day jaunt.

I believe the other contributors to this issue will sufficiently cover air travel and long trips by car. We certainly covered these topics in CF Roundtable the last time this topic came up. We can all sympathize with each other about the hardships air travel places on us, especially when we’re hand-carrying our precious medical equipment and refrigerated medications through security, to say nothing of oxygen or in-air emergencies. I suspect we all smirk a little bit when normal, healthy people complain about their bottle of water being confiscated, and secretly dub them a little pathetic. We are CFers and we know how things work and how to push back when we have to. Come to think of it, traveling with CF through airports has made me a much smarter, more prepared passenger than most of the people around me. These days, armed with printouts of what is allowed (from the TSA website and their new blog), I practically skate through security, even with my compressors and thermal of iced medications. Sometimes I’m forced to check my bag, especially if I’m in a Colistin month and am carrying needles – there are some rules not even medical necessity can trump.

So what about my time off? Well, this is where the motorcycle comes in. I love to take long trips and live out of my saddlebags and I am fascinated by the northern regions, especially the idea of going to Prudhoe Bay, on the north coast of Alaska. But how can a guy with CF reasonably make such a trip? A week of travel is no problem; most of us can do that. But going to Prudhoe would take far more than that.

Sometimes, I have to just stop thinking about the problem and do something about it. Last summer I joined six other riders on my longest, most challenging motorcycle ride yet: a total of 40 days of riding that took me all the way to the Arctic Circle in Alaska and the easternmost point of the United States in Maine. And through all of that, I kept up my therapies and maintenance faithfully.

Preparation is the key, of course. How could I keep refrigerated medicine cold on a bike for over a month? How could I even carry that much? I laid out all my medicines and stripped them of their packaging, including trimming off the extra plastic from the Pulmozyme® vials. I looked at the volume of 40 days of TOBI® and Pulmozyme and decided my current quart-size thermos wasn’t going to be enough. Luckily, Stanley sells a 2-quart size thermos on their website – I’ve never seen it in stores anywhere. Pack the medicine in that, fill it to the brim with ice, and tuck it away in my T-bag (the bag that sits on the passenger pad and acts as my backrest while riding). The other medications could be squirreled away in various other nooks and crannies, including pockets of my riding suit.

My compressor was a problem. I’d already lost one PARI Ultra® compressor while riding, when a saddlebag just plain fell off somewhere south of Syracuse. After that, I’d started carrying my medical gear only in the T-bag – but the regular home compressors are big – and require power. I spied a solution in a National Allergy flier: a portable, battery-powered compressor, the PARI Trek-S®. It was expensive, but worth every penny. While one may think of Alaska (and most of Canada) as a remote, barren wilderness with no running water or power; that accounts for only 99.99% of the land mass. The other .01% is along the roads – there are hotels, campgrounds, restaurants, everything. Everybody has running water, everybody has power. A few nights in some campgrounds, there were neither, but the battery of the compressor would last me through two treatments, and I always carried a lot of bottled water.

The Vest® was another problem altogether. It boiled down to the fact that I just couldn’t take it on the bike. Not only is it just too big, but the bike would probably vibrate it to pieces and destroy it – not something I’m willing to risk with something so expensive. With my doctor’s permission and a plan of alternative therapy in hand, I left the Vest at home. I rarely leave my Vest at home. I believe it to be an important part of my therapy and am very compliant in its use. But when traveling by motorcycle, one must make some hard choices sometimes.

A big part of the plan of alternative therapy was exercise. I have been a regular runner since October of 2004. While running hasn’t completely prevented exacerbations, I do believe it has helped lessen their severity, has held them off as long as possible, and has helped me recover faster and more completely. While traveling last summer, my left saddlebag held my running clothes and shoes. It often was great stress relief to my muscles to throw my bags into a hotel room or set up camp, then go for a short run of two or three miles. Once a week, I’d do a five or six mile run, to really help clear out the lungs. In fact, I felt better on the trip than I did most of the rest of the year at home.

A small part of the plan was to

Continued on page 23
By Maggie Sheehan

As I groan, huffing and puffing, pulling my Vest® up two flights of stairs almost every weekend, I ask myself two questions. Why do I go home so often and put myself through this agony of lifting my luggage and machines, and second, why do people give the heavy machines to the people who have the most breathing problems? Well, the first question is easy to answer. I should just stop going home so often. The second question I might have to ask a professional about.

These past two years in college I have realized I try to pack less and less every time I go home so I do not need to make a million trips up and down my apartment stairs. I saw my roommate come back from visiting home one weekend and I asked her where all of her “stuff” was? She proceeded to tell me that it all fit in her backpack. I was so shocked that she had one small backpack for three days. I carry home at least two duffle bags every weekend, and sometimes a third for my laundry. Forget packing frivously. I have to make sure I pack concisely and roll my clothes so small to make sure my PARI® nebs fit, my pills fit, and my Scandishake® cup fits. This bag does not include my Vest, of course, because it comes in its own carrying case. I am fortunate enough to now have two compressors, one for my parents’ house and one for my apartment at school. That at least conserves a bit of energy (and space for the laundry!)

Traveling with CF is not fun! I am sure we all have airport stories about people stopping us in security lines, but I wonder what has happened to common courtesy when it comes to seeing people struggling? A couple of years ago I was taking my first flight, ever, by myself. I was already apprehensive about being by myself on a plane, but thoughts whirled through my head about how I was going to get my Vest on the plane by myself and where I was going to store it. (This was before the carrying case was available on wheels!)

I struggled breathlessly down the center of the plane bumping every person on the aisle seats with my Vest. I was close to my seat when I finally found an opening in the overhead compartment. I used every ounce of energy my 5 foot, 92 pound body could muster to lift that Vest over my head. I got it half way to its destination when an older woman tapped me on the shoulder and proceeded to tell me I was blocking her way to her seat! I wanted to ask this rude, clueless passenger, “Excuse me! Do you not see me struggling with this heavy luggage?” But at that time I was a little shy and a lot less bold. I put my Vest down and let her through. I looked around and no one seemed willing to help me nor was there a flight attendant in sight.

I realized there was no way I was going to be able to lift this over my head alone so I tried to shove it under my seat! It really did not fit well under there, but no one seemed to care. That was the end of bringing my Vest on a plane. From then on I’ve decided to take my chances and check it with my regular luggage.

My boyfriend says he refuses to travel with me because he knows he would be carrying my Vest everywhere we went! I do not find that to be a bad thing...he apparently does! So what do I do about carrying heavy machines? Do I ask random strangers to lug my Vest up the stairs or lift it into my car? Actually, I know it is good exercise for me when I’m able, but I sure like it when my mom, dad, roommate, or a passerby happens to offer a hand! Who knows, with technology and everything else getting smaller, maybe someday I’ll be able to tuck it in my back pocket with my cell phone and iPod®!

Maggie is 19 and has CF. She is a Director of USACFA. Her contact information is on page 2.
Bullsh**ting

Hey, what’s the deal with your fingers?

You know, I been playing the horn for 36 years, man – That’s a lot of practicing. Flattens your fingers, man… heh heh.

– Scott Petersen
March 2006

"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...

DEBBIE AND LOUIE AJINI WITH THE BARENAKED LADIES AT THE 2008 NORTH AMERICAN INTERNATIONAL AUTO SHOW CHARITY PREVIEW IN DETROIT, MI.

ANA STENZEL AND ISA STENZEL BYRNES IN PORTLAND, OR, FOR ONE OF THEIR BOOK SIGNINGS FOR ‘THE POWER OF TWO’.

CRIS DOPHER AT THE ARCTIC CIRCLE, 200 MILES NORTH OF FAIRBANKS, AK, WHERE HE CAMPED OVERNIGHT BEFORE HEADING FOR HOME.
“I live where the river meets the sea.
I live where the waters of the Hudson end their cold journey to the Atlantic. My windows open to the East, the direction the Native Americans believe one must face to ask blessings for a new beginning. The morning sun glances off the water strongly enough to blind you. Late afternoon turns the entire West side of Manhattan to flame. At night the rising moon hangs above the jeweled horizon like the Eye of God. The river flows by me, and flows through me. Memory flows through me.

“I live where the river meets the sea, where push comes to shove, where love and anguish, blame and forgiveness, laughter and sorrow converge on the way to understanding.”

Thus begins Heather Summerhayes Cariou’s forthright and poignant account of her family’s journey with cystic fibrosis in “Sixtyfive Roses A Sister’s Memoir”. It’s beautifully written and integrates the present with the past in a most intriguing manner.

“Sixtyfive Roses” provides insight into a family’s life and how it is affected by CF. It tells of the good, the bad, and the ugly that is encountered during the family’s roller-coaster ride of a lifetime.

“Sixtyfive Roses” begins with the diagnosis of Pam, Heather’s sister, at the age of 4 and continues until the present. Heather makes a vow, at the tender age of 6, that she will be Pam’s protector and that they will die together. And then she spends the remainder of Pam’s life attempting to fulfill that promise.

Unlike many other books that are written about the suffering of a loved one, Heather does not attempt to canonize her sister nor present her family as the ideal family. While Heather and Pam are the best of friends, they still fight, as all siblings do. Heather resents all the time and attention that Pam receives, which leads to feelings of guilt. These emotions are the basis of many of Heather’s decisions regarding her own life and the direction that it takes. Many of Heather’s choices are somewhat self-destructive and extremely hurtful to her loved ones, as she acts out her frustrations, resentments, pain, and anguish. But through it all, the love and commitment of family are apparent.

Heather recounts the effects of CF on Pam, as well as her family’s responses, in a completely direct and honest portrayal. She describes how CF ravages Pam’s body, but never her soul. What Heather so vividly expresses is the havoc that CF inflicts on each family member. The toll it takes on her mother, who feels fully responsible for keeping the family running, no matter what obstacles are put in her way. She is a perfectionist who puts pressure on herself to be the “perfect” wife and mother while tending to the needs of Pam, whether at home or in the hospital. She expects that the rest of the family will handle things as she does – stoically and undeniably.

Heather’s father finds solace in forming the Canadian CF Foundation, which takes time, money, and energy away from his own family. And Heather’s two younger brothers bear the burden of a family in turmoil. This is especially so with the youngest, who also has CF and attempts to maintain both a physical and emotional distance from Pam.

Pam, herself, struggles not only with cystic fibrosis, but with her own aspirations and longings. She is a fighter, who demands independence and respect for her decisions. Pam is not about “giving up” but does understand that at some point she must surrender to that which befalls her.

“Now I live where the river meets the sea, married to the love of my life – a man who by his mere presence blesses me every day. Because I have loved and lost, because I love now, and because I know that loss is part of life, the taste of truth is bittersweet: there is no happy ending.

“But there is the day. The sun, the rain. The chance to say I love you. The willingness to forgive. The courage to remember. The opportunity to be kind. The ability to laugh and to be generous. The fact that we can choose our joy in each moment, no matter what. This, in itself, is the miracle.

“Pam said, tell our story. Mother said, tell the truth. The story I have told lies somewhere between truth and memory. Pam survives through the telling. So do I.”

Thus ends the tribute to not only Pam, but to the family whose belief system made them fighters and survivors of all that life has dealt them. ▲
Among the Guest Speakers:

Richard H. Simon, MD, University of Michigan, Ann Arbor, MI
“Improving Lung Function By Controlling Infection”

Dana S. Hardin, MD, Nationwide Children’s Hospital, Columbus, OH
“Cystic Fibrosis Related Diabetes”

Mark R. Elkins, PhD, Royal Prince Alfred Hospital, Sydney, Australia
“Strategies to Reduce Time For Airway Clearance”

John D. Mark, MD, Lucile Packard Children’s Hospital, Stanford, CA
“Complementary and Alternative Medicine”

Marybeth Howard, PhD, University of California San Francisco, CA
“Cystic Fibrosis Research”

Jonathan Widdicombe, PhD, University of California Davis, CA
“CFRI Research Dollars At Work”

Anna O. Tsang, RN, NP, MSN, St. Michael’s Hospital, Toronto, Canada
“Sexuality and Reproduction in Cystic Fibrosis”

Joseph Solowiejczyk, RN, MSW, CDE, Animas Corporation, Los Altos, CA
“Compliance”

Tiffany Christensen, Author and Patient Advocate with CF, North Carolina
“SickGirl Speaks!”

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In collaboration with United States Adult Cystic Fibrosis Association (USACFA)
Publishers of CF Roundtable

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Regular Registration after 7/1/08: $175 per person (includes meals)
Scholarships are available but limited for eligible applicants.

Cystic Fibrosis Research, Inc.
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CYSTIC FIBROSIS TEEN & ADULT DAY RETREAT

August 3–August 8, 2008
Located at Vallombrosa Center in Menlo Park, California

THIS YEAR’S THEME:

CF Club Med: Retreat of a Lifetime

Meet Some Great Friends! • Feel Like You’re Not Alone!
Learn more about taking care of your CF! • This is a place for hope and healing!

Who Can Come: Teens and adults 15 years and older with cystic fibrosis, their family members, friends and health care providers

Purpose of the Day Retreat: The retreat provides a safe and welcoming environment aimed at enhancing positive coping skills, social support and education for people who share common experiences with CF.

What We Do: Activities that promote health include daily exercise, arts and crafts, rap sessions, and educational workshops with guest speakers. Fun group-bonding activities include a talent show, games, and just hanging out getting to know others.

Cost: $65 per person for the entire week. Daily fees are $15 per day for visitors or $10 per meal for those who drop in for a meal only. Overnight accommodations and transportation are the responsibility of participants. Scholarships are available for those unable to pay fees.

Safety: All people with CF are required to comply with cross infection behavioral precautions. A medical advisor is available at all times, and volunteers are available to assist with respiratory treatments. Participants with CF must obtain a sputum culture before the start of the retreat.

People who have ever cultured Burkholderia cepacia, cultured Methicillin-resistant Staphylococcus aureus (MRSA) within the past 2 years, or are currently resistant to all antibiotics will not be allowed to attend the retreat.

We'd Love To See You There!
For An Application, Please Contact:

Cystic Fibrosis Research, Inc.
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Phone: (650) 404-9975/ cfri@cfri.org/ Fax: (650) 404-9981/ www.cfri.org
As I sit to write this article, I have just returned home from a successful vacation in Florida. I write ‘successful’ because I did not get sick while traveling and had a wonderful time. I find that being prepared and taking certain precautions or preparations for a trip helps keep one well enough to enjoy the vacation without an illness. That also means lots of lists, if you are forgetful like I am, and bringing enough medications plus some extra.

As much as I like to get away, the thought of plane travel is stressful for me. Mostly it is the thought of getting a cold from another passenger or sinus infection from the ‘bugs’ floating in the air. Another issue is that when I fly, I tend to get bad edema, especially in my feet, ankles and legs. I also worry I will forget important medications or apparatuses for delivery of medications. Lists become very important, and then checking them is imperative.

Since a lot of my medication needs refrigeration and I take it often, some of it, like Lantus and TOBI, might be the last thing I pack or forget to pack; as well as nebulizers and syringes. So, I check my list as I stuff my suitcase with clothes, toiletries and shoes but put a last-item check-list on the door handle. As I exit, I am reminded of stuff I could and have easily forgotten. This is minor but needs stating: never pack your medications in your suitcase. Always put important items in a carry-on bag due to possible loss of your checked luggage.

One strategy to staying healthy is to stay hydrated before, during and after a flight. I generally drink the recommended eight glasses of water a day and then some. Then the night before a flight, I try not to eat a salty dinner, or whatever meal, before I depart. This is hard as it means most of the foods that I love are off limits. Then I try to drink as much water as I can before I get to the airport. It is trickier and more expensive to drink a lot of fluids on the plane with the new travel restrictions before getting past security. I used to bring a two-liter water bottle with me on the flight from home. Now, I have to purchase all my water at the airport. Good for them, not for me.

The reason for all this water consumption is two-fold, I feel. First of all, water will act like a diuretic if you drink enough; meaning, if your body has plenty of fluid in it, it will not hold on to it and you will get less edema. It may be annoying to the person you have to disrupt to use the restroom but better on your body to drink a lot. Secondly, plane travel is very drying to your sinuses and lungs. Water will help to thin your mucus. Since there is less oxygen in a compressed plane it becomes very dry; it then becomes easier to cough stuff out if your mucus stays patent.

One thing to help prevent edema is to get up and walk around the plane. This is probably not encouraged by the flight attendants, but most doctors suggest it. Get up to use the lavatory. Or if the plane is set up with an aisle in the middle, do laps. On your walk, touch your toes, stretch for the ceiling.

Usually, before I travel, whether it is staying at someone’s house or a hotel, I make sure there is an exercise machine or gym that I can use when I land, or the next day. That helps get my circulation going and helps get rid of the collected swelling in my lower extremities. I also try to fit in a nap or a good night’s sleep after travel; that helps immensely to reduce swelling and increase peace of mind so you can enjoy your visit.

Once off the plane I try to get to my hotel or to the house of the person I am visiting and do a nasal lavage with a very portable plastic NETI pot, or with a WaterPik, to rinse out any possible bacterial particles I might have picked up on the plane. Some doctors and other CF friends have suggested wearing a surgical mask while seated to prevent possible infections. I find the masks to be difficult to breathe with and they usually aggravate my sinuses. Unless I am sitting next to someone coughing their guts out, I usually

“I try to gauge what meds I will need for the time away and then take an extra week’s worth.”
accept that I wouldn’t be able to get in every run and every treatment. I’d try my hardest, but travel requires compromise. The reality that I may be compromising my health was not lost on me. But I was feeling good at the time and that was just another reason to go; what if I never felt well enough again to do something like this?

I also took a little time to scope out the CF centers closest to my intended routes. I don’t often do this, but I figure if I got really sick, I’d need to go where they knew how to treat me. I should have also gotten fresh prescriptions from my doctor and taken those with me, in case I lost my medications, but I didn’t. Thankfully, I didn’t have a problem, but that flaw in my planning has stayed with me and won’t happen again.

So, equipped with plans, equipment, knowledge, crossed fingers, and two shiny new tires on my motorcycle, I took off. I’ll keep the travelogue brief. I met the group in Akron, OH, and we crossed Lake Michigan on the SS Badger. We drove across the top of the United States, through some of the flattest, hottest, most boring terrain I’ve ever been through (cows and corn and wheat and pigs). We finally got to the Rockies and headed through Glacier National Park, which is a destination itself. (Go before global warming destroys what’s left of the glaciers!) From there, we headed north, crossed into Canada, and joined up with Canada 2, which is the only land route through the Arctic Circle. It’s about a day’s drive from Fairbanks to get there and there isn’t much there, just some improved campsites and a marker declaring the latitude: 66° 33’ north. That is the line where, on June 21, the sun never sets, and on December 21, the sun never rises. To some of us in the contiguous 48 states, this is a magical demarcation, separating the real from the unreal, the normal cycle of night and day from a supernatural suspension of time. And though I was three weeks too late for a full 24 hours of sunshine, it never did get dark up there. I was awakened at 1 a.m. by the sun shining through the trees, through the window of my tent, and into my eyes. I got home. I had less than a day of turnaround time before the second half of the trip that took me to Eastport, ME. Laundry got done, of course, but so did a thorough examination of my remaining medications. I supplemented or replaced as necessary, packed up, and was off again. By this point, traveling on my bike, taking care of myself through twice-daily breathing treatments and every-other-day runs, was easy. My new travel partners were amazed, though, and looking back on it, I also am amazed at how disciplined I became – because I had to be.

I have to go back someday. I didn’t get as far north as I wanted, didn’t tempt fate enough, some would say. I know now, though, that what limited me then and might limit me in the future isn’t my CF – it’s time. I just ran out of time. The CF is manageable, with the right planning and equipment. The treatments are a known factor – there is no guesswork about those. Traveling with CF is always a question mark, but only because the rest of the equation is full of uncontrollable variables. But taking care of myself on the road? Done deal.

Andrea is 43 and is a Director of USACFA. She is the Executive Editor of CF Roundtable and Web master of cfroundtable.com. Her contact information is on page 2.

Cris Dopher, 36, is a lighting and scenic designer living and working in New York City. He has written one other article for CF Roundtable and runs to raise money for Team Boomer.
My Madness

By Kurt Robinson

While I am usually on top of it and have my article submitted in plenty of time, this article decided to catch the last flight out of town. The term “March Madness” took on a whole new meaning for me this year. Year in and out, March is a distracting month for one simple reason: basketball. It’s tourney time and there’s almost nothing that I’d rather be doing than sitting in front of a TV or in a gym watching hoops. Maybe that’s why this article wasn’t finalized until the last possible minute.

Looking back at my life, I have traveled more miles than I can ever begin to estimate watching, coaching, and playing the sport. I’m not sure why I love it so much because if you think about it, the concept is simple: put the round ball through the round rim. Whatever the case may be, I’m hooked and I’ve always been hooked! I have watched a lot of games that I can only equate to taking a nap (like the one I’m watching now) and have been witness to some of the best games ever (Duke vs. Kentucky, 1992 NCAA East Regional Finals). The tears have streamed down my face after a long and hard season when I was coaching, but I have also jumped for joy and pumped my fist in excitement after draining a long game-winning three-point shot my junior year in high school.

I have been able to travel throughout many western U.S. states for the purpose of something basketball related. When I was younger I was often on the court. As I’ve grown older, I’ve become more of a fan and enjoy watching the game even if I’m not necessarily rooting for either team. Coaching taught me to become more of a student of the game and try to analyze everything that is happening on the court. That being said, I still very much so get wrapped up in the excitement of a game. I am an emotional person and those emotions have always carried over onto the hardwood.

I was fortunate to have my dad coach me for many years. His passion for the game is like mine and his knowledge is even greater. We were able to share many memories together. As I grew older and my dad was no longer my coach, he always was at my games to support me and offer me advice and encouragement. I’m fortunate to have parents who have been such strong supporters throughout my life.

I know that basketball was key to my health remaining good. I believe that basketball is one of the best forms of exercise one can engage in. The constant jumping, running, springing, lateral movement, and quick thinking involved make for a good workout and a way to keep the lungs and body healthy. Through my senior year in high school I was able to play an entire game, practice five days a week, and travel, without my health being compromised. That was several years ago and in that time my health has gone somewhat downhill. It’s a tough truth to swallow that I am no longer able to go all out anytime I want.

Basketball has always been a release for me when I’m frustrated. I believe one reason that I enjoy basketball like I do is that, for the two hours or so I’m at a game, I can forget about everything else going on in life. This includes CF. I think it is important to have something in your life that you can turn to or engage yourself in that allows you to forget about life’s problems and stresses.

As I write this, the NCAA tournament brackets are only hours away from being announced and I can’t wait to fill out my bracket and watch hours and hours more of basketball over the few weeks that the tourney takes place. So with that I’m going to sign off and hope only that Duke will take home the trophy. Go Blue Devils! Take care and keep smiling! –Kurt

Kurt is 24 and has CF. He is a Director of USACFA. His contact information is on page 2.

In 1990, when Lisa McDonough was looking for people to continue publishing a newsletter for adults that have CF, Ken was one of four people who called her. He worked with the other three to create USACFA, Inc. He was the first Secretary of the organization and later became Editor.

Ken served as Editor of CF Roundtable from the second issue until January 1993. He had definite ideas of what he thought CF Roundtable should be and worked to try to get those ideas adopted. He solicited articles from many sources and enjoyed his editing chores. He was a lifelong resident of Illinois, and had worked on various newspapers, including the Chicago Tribune, the Register-Mail in Galesburg, Illinois, and the Joliet Herald News.

Ken never married but he enjoyed being “Uncle Ken”. He was very proud of his nephew and nieces. He wrote about them in CF Roundtable Autumn 2002.

People in the Chicago area will remember him from The Chosen Few and CF Awareness Days of the past. He was a regular attendee and enjoyed the contacts and friends he made in those groups.

He always felt that the future was bright for adults that have CF and that a cure for CF was “just around the corner”. He liked to end his articles with a cheerful note. To honor Ken we end this with his tagline: “Until next time, take care.”

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**Benefactors**

**BRONZE**

Colleen Adamson  
Blanche & Derwin Ball  
(in memory of daughter, Marsha Ball-Waldo)  
Isabel Stenzel Byrnes  
Dorothy Corwin  
(in honor of son, Raymond R. Corwin on his 11th transplant anniversary & his 47th birthday)  
Paul DeSalvo  
Mr. & Mrs. Werner Freundlich  
David Henley  
John & Joanne Jacoby  
Janet King  
The McLaughlin Family

**GOLD**

Anonymous  
(in honor of Pammie Post)

**PLATINUM**

CF Services Pharmacy  
Maggie’s Miracle Makers

Those who give $50 or more in a year will be recognized as Benefactors, unless they request anonymity. The categories are: $50-$249 Bronze Benefactor, $250-$499 Silver Benefactor, $500-$999 Gold Benefactor, $1,000 and over Platinum Benefactor. Donations over $10 are tax deductible. Please make checks payable to USACFA, Inc. Send donations to: USACFA, Inc., PO Box 1618, Gresham OR 97030-0519
Transplant recipients are at a greater risk of developing skin cancer due to the immunosuppressants they take. I have learned this the hard way, having had multiple pre-cancerous spots on my hands, neck, face and scalp scraped and burned off. I have also had two major surgeries for skin cancer, one surgery on my nose and one over my right eyebrow.

I am now preparing for my third major surgery for skin cancer, this time on my head. It's important to note here that my dermatologist had been treating my scalp by scraping off the spots which were precancerous at the time. One spot did not heal all the way which is an indication that the spot had turned cancerous, and off I went to the dermasurgeon. You know it's bad when you go to see the dermasurgeon and he says, “Oh my” when he looks at your head, and then asks for a ruler! He measured a 9x5 cm patch of skin cancer on my head that needed to be surgically removed. He also talked about reconstructive surgery and plastic surgeons, which is hard to hear, because then you know it’s especially bad.

I will have the dermasurgery one day, and I will have reconstructive surgery the next day to put a grafted piece of skin from my leg onto my head. At least they aren’t taking the skin from my butt because then I would literally be a butt-head! I will also have skin extenders under my skin, which are essentially saline balloons they will put under the good skin on both sides of the graft, and gradually add saline to keep extending the skin. Eventually the extended skin will be large enough that the plastic surgeon can use it to cover my bald spot (the grafted area). Pretty cool, huh? This is a four-month process which will have me looking like Mickey Mouse with the saline balloons on my head resembling mouse ears!

So now I am trying to get used to that idea. I already told my boss I won’t be going to any meetings for the next four months! Seriously, though, the way I am dealing with this is to describe to everyone what I will look like so they are prepared for it. I feel better knowing that people won’t be surprised by my appearance, and knowing that I won’t be stared at too much. I am thinking about wearing scarves on my head so people can’t see what’s going on on the top of my head. I still haven’t decided what to do about that – leave it exposed or hide it. I’m usually very open about my health issues so I may decide to leave it exposed and treat it as an educational display! Wear hats, people!

People actually think that I did this to myself by being out in the sun without sunscreen or a hat. Plus I am fair-skinned. However, this has little or

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**Calling All Writers**

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

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

Here are a few possible topics for your use: headaches; understanding what you hear; pain(s) in the neck; arm twisting; the case at hand; a breath of fresh air; gut reaction(s); pain in the butt; oh, my aching back; getting hip to a subject; standing on one’s own two legs; at the foot of the problem; toeing the line; my sole responsibility. As you can see, these are humorous suggestions that are meant to give you some ideas. You need not use any of these, but you may, if you wish. For other ideas, check out the Looking Ahead section on page 5. All submission dates for the coming year are posted there.

We ask that all submissions be typewritten. If you want to e-mail your submission, please have it in Microsoft Word or a similar program. You may send your submissions to: cfroundtable.com or to

USACFA
PO Box 1618
Gresham, OR 97030-0519.
nothing to do with my skin cancer issues. I always have sunscreen and a hat on when I go outside. I treat these encounters as educational opportunities and tell them it’s really because of the immunosuppression medications that I take and that people with transplants are, in general, more susceptible to skin cancers. Many people with transplants don’t even realize this and are not necessarily told this by their transplant centers. People with transplants really need to see a dermatologist regularly and be examined for any evidence of pre-cancerous lesions or cancerous spots. As one of my transplant coordinators said, the farther out from transplant you are, the more regularly you should see your dermatologist, and that you should see your dermatologist more often than the transplant doctors!

Another thing I have learned from these experiences with skin cancer is people with transplants really should, if possible, see a dermatologist who is familiar with transplant patients and their skin cancer issues. My first dermatologist didn’t have this experience and, therefore, he didn’t take some of the spots I had on my face as seriously as he should have. There was one spot that he downplayed the severity of and it was then up to me to call the dermasurgeon to have it removed. By the time I did that, the skin cancer had advanced deep into the skin and I had to have a lot of skin removed (including most of my eyebrow!) to get it all out. It was a very scary time for me, and I partially blamed myself for not realizing the severity of the problem early enough. I had trusted my dermatologist and he had failed me miserably. That is when I changed dermatologists and I now see one that has a lot of experience with transplant patients. I have a lot of faith in him because he doesn’t let things go far before he treats them or has me see the dermasurgeon.

Please learn from my experiences. If you have had a transplant, and you have not seen a dermatologist, please get a referral from your transplant team for a dermatologist who sees transplant patients, and make an appointment! If you have spots that are scaly and won’t go away, get them checked out. Skin cancer is a very serious issue, and if left unchecked it can cause a great deal of problems for you. You need to keep on top of this, but remember even if you do keep on top of it you can still have problems as I have illustrated. However, the problems will be easier to deal with if you have a good dermatologist by your side.

Colleen is 39 and has CF. She had bilateral lung transplant on July 3, 1998 and a kidney transplant on March 7, 2006. She is a Director of USACFA. Her contact information is on page 2.
A DEEP BREATH IN
Setting Limits and Pushing Boundaries

Debbie Ajini

CF has evolved over the years on so many levels. Back in the day, kids with CF were encouraged to sit down and watch. They were told to “take it easy”. I remember getting out of many gym classes due to my CF. The funny part is my lung function was probably the best then and I was encouraged not to do too much. Now we know starting exercise at an early age is one of the best things someone with CF can do.

I guess as with any culture or group, you evolve. We have learned so much about CF in the past 30-40 years that things are approached very differently today than they were when I was a kid. Now I enjoy pushing the boundaries of what someone with CF should or shouldn’t do. And as my lung function has declined, I have learned to set limits for certain things.

One aspect of setting limits that I am struggling with is travel. I have written in a few columns about traveling. My husband and I enjoy it. We haven’t been to that many places in the world, but anywhere with a beach is a good start. As my lung function has declined, so have our travel plans. As I am sure others will share, it takes some effort to plan for a trip for someone with CF. Planning takes more work for me now that I use O2. As I wrote in the Spring 2006 and Autumn 2007 issues, I have really worked on figuring out the logistics as far as setting up O2 delivery, what to pack etc. Then if you throw in O2 and things like the Bi-pap or tube feedings it adds to the planning.

Not to mention the actual work of being on vacation! I know that, to healthy people, sounds silly. But to those with CF, we know it takes a toll. Traveling wears down even the healthiest person. I certainly do not need a new bug to fight. Coincidentally, the major crisis for me began the day before I left for Mexico. I took an oral antibiotic with me. I felt OK but did spike a few fevers. Over the next two months things progressed from orals, to IVs, to the hospital, to the ER. I still wonder if that vacation made whatever was starting worse. Yet, if it did, I don’t regret going.

Our last vacation was over a year ago. On the cruise I did not use a wheelchair. I walked all day. A lot of the time my husband carried my O2 but I walked. And I felt great. So, I know I could probably push my boundaries more. Part of me really wants to go somewhere. I think a warm, sunny place would be a good boost for me. I know my husband could use the break too. But I just don’t know. I feel better just doing trips here in Michigan and being close to my home base.

We have dreamed of going to Fiji for many years. Do we not go because of the lack of healthcare and knowledge of CF, or do we plan as much as we can and go? Do I travel at all? Then, if so, how far from home am I willing to go? Or, more specifically, how far from a CF center do I go? I worry that I am limiting myself unnecessarily. The mountains in Colorado will be just as breathtaking this week as in 10 years from now. The ocean in Fiji will be just as blue. But I am scared. Scared to get out of my safe zone. As I get closer to the process of transplant, I do not want to mess anything up.

I just found out I am going to Pittsburgh in July for a transplant evaluation. Once I cross that hurdle, part of me feels like we should travel then. I have a hard time with the phrase “After transplant...” On the one hand (see, I need more hands!) I understand that right now there are things I simply cannot do that I will be able to do “After transplant...”. But then there are things like having a big party, redecorating a room in our house, or traveling that I really could do right now. It would just take more time to plan and eventually execute. So I wonder is it worth it for my mind to do these things now or to wait? What is the balance between me setting my limits and pushing the boundaries?

I do know it is not healthy, mentally or physically, to sit back and not participate in life. The challenge for me now is how much should I do, how much can I really do and how will I feel after I do it? I may have many more limits that I set on things, but I also know there are still a few boundaries I can push! 

Debbie is 37 and has CF. She is a Director of USACF. She and her husband, Louie, share their home with their yellow lab, Max. Her contact information is on page 2.
Nominate Your Everyday Hero

Do you know any unsung heroes with CF? At USACFA, we continue to be amazed at the achievements and attitudes of many adults in the CF community. For nearly four years, Genentech’s Heroes of Hope Living with CF program has recognized people with CF who exemplify heroism. Some of the past heroes inspired people through their music, provided thousands of people with free CF legal advice and volunteered in their own local communities. Heroes of Hope award recipients are united by their common traits of maintaining a positive attitude, proactively maintaining their health and making the most of their lives. We know that there are many more everyday heroes out there in the CF community waiting to be nominated.

As some of you may know, Heroes of Hope has recognized almost 40 outstanding individuals with CF across more than 30 states. Anyone can nominate a person with CF to be a Hero of Hope – just log onto the newly-renovated Heroes of Hope Web site, www.heroesofhope.gene.com, to download and fill out a nomination form, and fax or mail it in. An independent Heroes of Hope Advisory Panel comprised of seven members of the CF community regularly selects nominees to be Heroes of Hope recipients. In the past, Heroes of Hope recipients were recognized in small ceremonies at their local CF center. This year, the program is moving online which will give everyone in the CF community access to the inspirational stories and advice of each Hero. Once selected as a Hero of Hope recipient, each will now receive the following distinction:

- Heroes of Hope recipients will be able to record a downloadable podcast
- A personalized page on the Heroes of Hope Web site for each recipient that will feature their podcast, a collection of their photographs and their biography
- Heroes of Hope recipients will receive an award, a certificate and a t-shirt
- Your story or that of your child will reach thousands of people in the CF community.

Do you know a Hero of Hope? To nominate that person (or several people!) and to learn more about past award recipients, please log onto <www.heroesofhope.gene.com> today.

GoodSearch.com Helps Raise Money For USACFA

Every time you use the internet, you can raise funds for USACFA. Go to: GoodSearch.com each time you want to surf the internet or find anything. They will pay us for every search by a user who designates USACFA as their charity of choice.

We just received another check from GoodSearch. Although it isn’t lots of money, every little bit helps. Just use GoodSearch when you search the internet. Designate USACFA as the charity of your choice, and we get a few cents for each time you use it. This is a painless way to contribute to USACFA and we appreciate the help.
Hello everyone, and welcome to another edition of “Unplugged”. Time surely does fly, doesn’t it? Lately I have been thinking about honesty, limits, and frustration with CF and, coincidentally, our person in the spotlight hits on those issues and more. This edition I turned “Unplugged” on its head.

Instead of interviewing an adult with CF, I sat down with someone who works with us. I thought it would be interesting to see things from a different angle, and to explore the CF experience from the other side. I wanted to find out what others think of me, I mean us, as a group. I interviewed Michelle Murray, Nurse Coordinator at California Pacific Medical Center. (Full disclosure: I go there for my care so I totally love her. I swear.)

So here is Michelle:

1. Name - Michelle Murray
2. Age - 42
3. Location – San Francisco, CA
4. When did you first start working with people with CF?
   July 2005. Prior to taking this job I worked in the Adult Medical-Surgical ICU and facilitated clinical research studies. I needed a change and the job as Nurse Coordinator for the Advanced Lung Disease Program was something different and challenging.
5. Where do you work?
   California Pacific Medical Center, Adult CF Center, Co-directed by Noreen Henig, MD and Christopher Brown, MD.
6. How would you describe the clinic? (Size, age range, health, etc)
   Approximately 40 adults aged 18-59. Patients travel to the clinic from the Oregon border, Central California, and western Nevada. I’d say 65% are in good health, 25% have good months and bad months, and 10% are really struggling with their health. This year we increased our outpatient multidisciplinary support and added an outpatient social worker and nutritionist in addition to the great inpatient nurses, social worker, and dietary team we’ve worked with for years.
7. What was your welcome to CF moment?
   When I was in seventh grade, a good friend was diagnosed with CF. I remember visiting her when she was in the hospital. At the time, I was quite intrigued by her PICC line.
8. How do you find people with CF to work with?
   People with CF have their own personalities, backgrounds, needs and interests. As individuals with different personalities and disease courses, people with CF keep my work life interesting, challenging, (occasionally frustrating), and fun. One of the best parts of my job is getting to know everyone as individuals.
9. Do you think we are more or less religious?
   Neither, I think religion is personal and not really connected to whether you have CF. Some of the CF patients I work with have found strength and comfort from religion and it is part of their support system.
10. What is the biggest hurdle for adults with CF? Is denial a big part?
    One of the biggest hurdles is juggling adult responsibilities (job, insurance, maintaining a home, raising a family, caring for aging parents, etc.) and finding time to care for oneself. Daily CF care is a burden. The trick is finding a manageable schedule that pays off with better health. Yes, I do see denial. I see it more when CF is progressing and when someone is having a hard time managing their health. That seems to be the time when people tend to deny what is really happening.
11. Do you see any evidence we have a hard time with relationships?
    Some people do have a harder time with relationships. But I also find that people with CF are frank and forthcoming. You either accept them for who they are, or move on because they have life to live.
12. Is there one thing we are afraid to talk about?
    I can’t think of any one topic the people with CF won’t discuss. In general, I think people with CF avoid talking about their treatment compliance or lack thereof. That’s human
nature. I’m not very good at flossing my teeth, and I’m not always completely truthful when I visit my dentist.

13. What do you love about CF?

The challenges it presents and the new and cutting edge scientific and clinical changes that are happening.

14. What do you not like so much about CF?

Honestly? The sputum.

15. What is the predominant attitude with us?

Your fighting spirit. You are survivors.

Sometimes I think I am unique, so different from most people, due to my CF. No one else has to do the Vest® or chest PT, or do as many sinus irrigations or bowel cleansings. Nor do they have to stand three feet away from their friends and resist the urge to give them hugs. But many people use the Nettie-Pot and take laxatives (maybe not as many). The truth is, everyone feels alone and isolated at times. Our isolation is more tangible and emotional. I don’t see people who understand not wanting to do the Vest. One theme I got from Michelle was that, yes, we have certain differences and unique challenges but there are many common experiences that make us similar to everyone else. It is all a matter of degree and perspective.

OK, I have lied to my doctor about compliance issues. Shocking! Yes, I have trouble balancing my life, my healthcare, my work, my other activities and relationships. And I know it is more challenging for us due to the amount of time we take to care for ourselves and manage our disease. And then there is denial. Yes, there have been times when I have been in denial about my CF. My denial takes all shapes and forms, from not talking about it with a partner, to not taking care of myself.

Once, when I was 18 and a freshman in college, I decided not to take my enzymes. I have no idea why, but it made sense at the time. Well, let’s just say it was not pretty. I think I spent more time in the bathroom and ruining my boxers than accomplishing my intended goal. Today I don’t do so much to sabotage my health, but denial is a convenient option. Thankfully, these days I am much more gentle with myself. While I tend to not want to burden people with my health issues or find it challenging to stick within my health regimen, if I do get off course I get back to taking care of myself much more quickly. I’ve learned to practice love for myself and acceptance of my CF. I think my denial arises out of a desire to not have it.

So, I don’t know about you, but I am confused by all this stuff. I am different and I am not different. I accept and struggle with my disease, yet I deny it all sometimes. I just want to be like my friends who get up for work and take a shower and leave. I don’t know how to put it all together, but I try not to beat myself up over it. The key for me is to know, and fulfill, my responsibilities. I have to take care of myself the best I can. And also to accept that’s it’s difficult, and I have feelings—lots of them—and they are not bad. I just cannot let them dictate how I care for myself. Today I know that I am a uniquely average adult with CF.

Rich is 39. He has CF and is a Director of USACFA. His contact information is on page 2.

TILLMAN continued from page 13

Therapeutics, Inc., (CFFT), the non-profit drug discovery and development affiliate of the Cystic Fibrosis Foundation, has selected Hollis-Eden’s TRIOLEX™ (HE3286) as a drug candidate for lung inflammation associated with cystic fibrosis (CF). TRIOLEX is a novel orally bio-available adrenal steroid hormone analogue with anti-inflammatory and insulin sensitizing properties currently in clinical trials under an open Investigational New Drug application (IND) for the treatment of metabolic disorders. CFFT selected TRIOLEX as a drug candidate for lung inflammation associated with CF based upon the potent anti-inflammatory activity and attractive safety profile of the compound in preclinical studies to date. In addition to its anti-inflammatory properties, TRIOLEX has also demonstrated in preclinical models glucose lowering and bone sparing activities that could provide added benefit to patients with CF.

Data Show PTC124 Addresses Underlying Cause of Genetic Disorders and Restores Protein Function in Cystic Fibrosis Model

PTC Therapeutics, Inc. today announced the publication of new preclinical data in the February 12, 2008 edition of the Proceedings of the National Academy of Sciences (PNAS) which show that PTC124, a novel drug designed to bypass nonsense mutations, was active in a preclinical model of cystic fibrosis (CF). These results support and add to research published last year in the journal Nature, which demonstrated the activity of PTC124 in a preclinical model of Duchenne muscular dystrophy (DMD). PTC124 has demonstrated pharmacodynamic proof of concept in
Phase 2a clinical trials in nonsense-mutation-mediated CF and DMD. PTC124 functions by overcoming the premature stop signal and reading through the complete genetic instructions, resulting in the restoration of a full-length, functional protein. Patients with CF lack the CFTR protein, a chloride channel that maintains proper hydration of epithelial cells in the lung, pancreas, and liver. The data in PNAS demonstrate that PTC124 allows CFTR to be made in cells in which it was previously absent, to be delivered to the proper cellular location, and to induce chloride channel function.

**Inspire Initiates Second Phase 3 Cystic Fibrosis Trial**

Inspire Pharmaceuticals, Inc. (NASDAQ: ISP) announced the initiation of TIGER-2, its second pivotal Phase 3 clinical trial with denufosol tetrasodium inhalation solution for the treatment of cystic fibrosis (CF). TIGER-2 (Transport of Ions to Generate Epithelial Rehydration) is a 24-week, double-blind, placebo-controlled, randomized study comparing 60 mg of denufosol inhaled three times daily to placebo in approximately 350 CF patients with FEV1 (Forced Expiratory Volume in one second) greater than or equal to 75% of predicted normal. The trial’s primary efficacy endpoint is change from baseline in FEV1 (liters) at the 24-week time-point. Secondary endpoints include other lung function parameters, pulmonary exacerbations, requirements for concomitant CF medications and quality of life. Based on pre-clinical and clinical work, denufosol, a selective P2Y2 receptor agonist, has several pharmacological actions contributing to its mechanism of action: hydration of the airways by stimulating chloride and liquid secretions on the epithelial cell surface; inhibition of epithelial sodium absorption; enhancement of ciliary beat frequency; and stimulation of mucin secretion. Denufosol for the treatment of cystic fibrosis has been granted Fast Track designation and orphan drug status in the United States by the FDA and orphan drug designation in Europe by the European Medicines Agency (EMEA).

**FDA Grants Orphan Drug Designation to Mpx for Levofloxacin Solution for Inhalation**

Mpx Pharmaceuticals, Inc. announced that the U.S. Food and Drug Administration’s (FDA) Office of Orphan Products Development has granted Mpx orphan drug designation for levofloxacin solution for inhalation for the treatment of pulmonary infections due to Pseudomonas aeruginosa and other bacteria in patients with cystic fibrosis. Levofloxacin is the active pharmaceutical ingredient in MP-376, the company’s proprietary levofloxacin solution for inhalation. MP-376 is a proprietary formulation of levofloxacin that has been designed by Mpx scientists to enable aerosol administration. Administration of MP-376 with a high efficiency nebulizer to the lungs allows for the delivery of high concentrations of active drug directly to the site of infection, while minimizing systemic exposure. Mpx believes this approach has the potential to improve bacterial killing and reduce resistance development versus traditional oral or IV routes of administration.

**ANTIBIOTICS AND THERAPIES**


Two multinational, double-blind, pivotal Phase 3 studies involving more than 1,600 patients have shown ceftobiprole to have in vitro activity against penicillin-resistant Streptococcus pneumoniae, Pseudomonas spp., and methicillin-resistant Staphylococcus aureus (MRSA). Adverse effects included nausea, vomiting, taste disturbance, headache, and diarrhea. Ceftobiprole is an essential addition to the antimicrobial armamentarium for use against MRSA and/or multidrug-resistant gram-negative infections. The safety profile is consistent with that of the cephalosporin class of antibiotics.


**New Pulmonary Therapies for Cystic Fibrosis.** Felix Ratjen, MD, PhD, FRCP©. Curr Opin Pulm Med 13 (6):541-546, 2007

This review addresses recent therapeutic strategies that either target the underlying defect or early steps in cystic fibrosis pathophysiology. While gene therapy does not appear to be a therapeutic option in the near future, cystic fibrosis transmembrane regulator pharmacotherapy is currently being developed as an alternative to reduce cystic fibrosis transmembrane regulator degradation or improve its function. Two drugs that increase chloride secretion via an alternative chloride channel, Moli1901 and denufosol, have been shown to be safe in clinical studies that also suggested clinical efficacy. Osmotic therapy may be an alternative approach to increase airway surface liquid and is being studied as an early intervention strategy. Currently it is too premature to conclude which one of these approaches will turn out to be most promising. It is rather likely, however, that at least one of them will make a major difference for patients and change the way we treat cystic fibrosis lung disease by moving from treatments that address the downstream effects of CFTR dysfunction to a more causal approach to treatment. While the prospect of therapies targeting early aspects of cystic fibrosis pathophysiology is certainly exciting,
Early diagnosis, treatments of acute exacerbations, and chronic therapies have all improved the lifespan of cystic fibrosis patients; however, the natural history remains one of worsening bronchiectasis and obstructive airways impairment. The progression of disease leads to eventual respiratory failure, but some will have other acute respiratory complications that require intervention. In this report, we review the most common life threatening respiratory complications of cystic fibrosis, including pneumonia, massive hemoptysis, and respiratory failure.

http://www.ajms.com/pt/re/ajms/abstract.00000441-200801000.0011.html;jsessionid=HwcbFyv5QFzxy4yQssCnYl8rFhdgnD20XdkgypkWcpNMb9xF!-809317659!181195629!8091!-1

Airway Inflammation in Cystic Fibrosis. Arnon Elizur, MD; Carolyn L. Cannon, PhD and Thomas W. Ferkol, MD. Chest. 2008; 133:489-495

Patients with cystic fibrosis (CF) experience declining pulmonary function related to chronic airway inflammation, which results from epithelial and immune cell secretion of proinflammatory mediators that promote neutrophil influx into the airways. This inflammatory response may be disproportionate to the inciting infectious stimulus, resulting in an overly exuberant influx of neutrophils. The neutrophils release proteases, including neutrophil elastase, that eventually overwhelm the antiprotease capacity of the lung and cleave structural proteins, leading to bronchiectasis. This deleterious inflammatory process in patients with CF has become a potential therapeutic target, though the development of effective antiinflammatory therapies has been limited by the lack of sensitive outcome measures. Historically, indirect measures of lung disease, such as spirometry, have been used to assess the effect of antiinflammatory drugs. BAL remains the primary method of interrogating the inflammatory status of the airway, but the procedure is invasive and may eventually be supplanted by induced sputum. Anatomic imaging with high-resolution CT scanning is used clinically, but has unknown utility, and functional imaging, using positron emission tomography, appears promising but is still investigational. Despite the paucity of outcome measures, clinical trials of antiinflammatory agents, including corticosteroids and ibuprofen, have demonstrated benefits, though their use has been limited by adverse effects. Azithromycin is increasingly used as an immunomodulatory agent, although its mechanism of action remains unclear. Strategies for modulating the airway inflammation in patients with CF are currently under development and may offer new therapeutic options for these patients.

http://www.chestjournal.org/cgi/content/abstract/133/2/489

Risk factors for increased need for intravenous antibiotics for pulmonary exacerbations in adult patients with cystic fibrosis. N. A. Jarad, K. Giles Chronic Respiratory Disease, Vol. 5, No. 1, 29-33 (2008)

This study investigates factors associated with pulmonary exacerbations (P Exs) in a large cohort of adolescent and adult patients. In this study, risk factors for P Exs were found to be as follows: growth of P. aeruginosa in the sputum, reduced FEV₁ and CF-related diabetes mellitus. Age, gender and BMI did not influence the annual number of exacerbations.

http://crd.sagepub.com/cgi/content/abstract/5/1/29

Laura Tillman, 60, has CF. She is a Director of USACFA and is the Secretary. Her contact information is on page 2.
The Boom er Esiason Foundation (BEF) is pleased to offer several different scholarship opportunities, available both annually and quarterly. In 2008, over $1,500,000 in scholarship money will be awarded.

Launched in 2003, the various scholarships are intended to assist people who have CF and are pursuing undergraduate and graduate degrees. The scholarships are awarded throughout the year and are based on a variety of factors, including: academic accomplishment, commitment to healthy living, and demonstrated financial need. The grants are made directly to the academic institutions to assist in covering the cost of tuition, and room and board.

One of the newest scholarships, from the University of Phoenix, presents an exciting opportunity to those with CF, who because of health limitations are unable to attend college physically, and have chosen to pursue an online degree. In 2008 the University of Phoenix is contributing 26 full-tuition scholarships to BEF for one degree program, each. University of Phoenix offers associate, bachelor, master, and doctorate programs.

BEF Scholarships are awarded quarterly with deadlines for applications on March 15, June 15, September 15 and December 15, 2008. The deadline for applying for the University of Phoenix scholarships was March 28, 2008. The Exercise for Life scholarship application deadline is June 27, 2008. The Bonnie Strangio scholarship applications are due by June 13, 2008. Applications for the Scholarship of the Arts are due by May 23, 2008.

Go to: www.cfscholarships.com for more information. Recipients of these scholarships all agree that it is easier to focus on academics and treatment, when finances are less of an issue. When you go to the BEF web site, you will find links to scholarships that are offered by some pharmaceutical companies.

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- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq.**, 811 Rusk Street, Suite 712, Houston, TX 77002-2807.
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IMPORTANT RESOURCES

For a directory of pharmaceutical companies that provide free prescription drugs to patients who qualify, write to: Directory Programs, Pharmaceutical Manufacturers Association, 1100 15th St. NW, Washington, DC 20005-1707. Use the information provided in that directory to contact the appropriate companies for information on their specific programs.

United Network For Organ Sharing (UNOS). Phone: 1-800-24-DONOR. Call for information on transplant centers, access for all patients needing organ transplants and general transplant information.


American Organ Transplant Association (AOTA): Helps defray out-of-pocket travel expenses for transplant recipients. Helps to set up trust funds. For more information write: American Organ Transplant Assn., 3335 Cartwright Rd., Missouri City, TX 77459-2548. Or call (281) 261-2682. e-mail: infoAOTA@a-o-t-a.org.

ADA: To learn how the Americans with Disabilities Act (ADA) applies to you, contact the Disability Rights Education and Defense Fund (DREDF) at 1-800-466-4232 between the hours of 9 a.m. and 3 p.m. Pacific Time. The DREDF has received a federal grant to inform individuals, who may be subject to discrimination, of their rights and remedies. To report a violation of the ADA, call the Equal Employment Opportunity Commission (EEOC) at 1-800-669-3362.