CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS AUTUMN 2014

Special Needs Trusts And SSI Benefit Eligibility

By Beth Sufian, JD & Amy Sopchak, JD

The *CF Roundtable* column "Ask the Attorney" has provided information to readers over the years regarding Social Security benefits for people with CF. Almost half of the CF population receives some sort of Social Security benefit. Approximately 60 percent of all adults over the age of 18 receive either Supplemental Security Income (SSI) benefits or Social Security Disability Insurance (SSDI) benefits.

A person who is eligible for SSDI benefits does not have to worry about how much he or his spouse has in assets. SSDI eligibility is not based on the assets of the applicant. If a person receives an SSDI check that is more than the amount of the SSI benefit in his state, then he is NOT eligible for SSI benefits.

For those individuals with CF who have not worked enough to receive SSDI benefits or whose SSDI check is less than the SSI amount in their state, SSI benefits may be an option. However, in order to be eligible for SSI benefits, the SSI recipient must meet certain income and asset eligibility criteria. In addition to meeting Social Security Administration (SSA) medical criteria, SSI eligibility is also based on the assets and the income of the applicant. If the applicant is married, the income and assets of a spouse are also counted. SSI is available only to individuals who have assets below $2,000 for an individual and $3,000 for a married couple or family of more than two people.

An adult with CF who is over 18 and living with his parents will not have the assets or income of his parents counted for purposes of SSI asset eligibility. However, if the adult with CF has his own assets that are over the SSI eligibility limit of $2,000 for an individual, the adult will not be eligible for SSI benefits. Even if a person meets the Social Security medical eligibility criteria, the person must also meet the SSI income and asset eligibility criteria.

> "Social Security will not count the assets in the Special Needs Trust when determining eligibility for SSI."

This article will address how a Special Needs Trust may help a person with CF qualify for SSI benefits even if the person has assets over the SSI asset eligibility criteria prior to applying for SSI benefits.

Continued on page 12

INSIDE THIS ISSUE

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EDITOR'S NOTES

Autumn has begun and, so far, is beautiful. It is time to talk with your health team about flu shots.

I'm sorry to report that **Jen Eisenman** has left the Board of Directors of USACFA. She served four years and was very active with social media and communicating with all the CF Adult Clinics in the country. We wish her well and will miss her. One other sad note is that **Jim Chlebda**, who authored "Creative Disengagement," has died. See his obituary on page 23.

I hope you have read the article about Special Needs Trusts and SSI Benefit Eligibility that starts on the front page. **Amy Sopchak** is a lawyer who has special experience working with people who have CF. In the same vein, **Mark Manginelli** discusses wills and trusts in "Protecting What Matters." Both of these articles blend with our Focus topic: Dealing With The Death Of A Loved One With CF.

I think this topic proved to be harder to write about than many people thought it would be. Although we have had requests from family members to print articles they have written about a loved one who has died, none of them wrote for this issue. Four of us did write about the death of a loved one. **Mark Levine** wrote of his late brother, **David**. In "Spirit Medicine," **Isabel Stenzel Byrnes** wrote of many friends and her beloved sister **Ana**. In "Speeding Past 50, I wrote of many dear friends. **Julie Desch** wrote of her sister, **Kathy**, and brother, **Tom**, in her column "Wellness."

On a happier note, we offer congratulations to Julie Desch on her receipt of the first ever **CFRI Partners in Living Award in Memory of Anabel Stenzel**. This award is given to a person who has CF, has supported CFRI through volunteer and/or fundraising efforts and embodies the qualities exemplified by Ana, including courage, initiative, determination, adherence to medical regimen, community service and positive coping. Well done, Julie, we're proud to know you.

Be sure to read our new column, "Parenting," written by **Karen Vega**. She hopes that many of you will contribute to the discussion.

Joan Finnegan Brooks, a former Director of USACFA, is the feature of "In the Spotlight." The "Conversation Corner" finds **Jenny Dolan** describing her "CF Table." **Angelo DiStefano** uses "Voices from the Roundtable" to tell of his stem-cell treatments. **Laura Mentch** tells us of two new airway clearance devices that she has used and finds effective. Once again, **Laura Tillman** has compiled a wonderful list of "Information from the Internet."

Jeanie Hanley tells of her experience at the CFRI Education Conference. Be sure to read of the bike adventure undertaken by **Jerry Cahill** and **Emily Schaller**. (See page 32.)

For those who have been wondering how the CF case in Arkansas is doing, check out "Ask the Attorney" where **Beth Sufian** explains about the case and the outcome.

Stay healthy and happy.

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MILESTONES

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

ANNIVERSARIES

Birthday

Michael Kolterman, 55
Monticello, MN
On September 7, 2014

Gina Pfau-Moriarty, 30
Spokane, WA
On January 10, 2014

Kathy Yoder, 52
Portland, OR
On August 16, 2014

Wedding

Michael and Caryn Kolterman
Monticello, MN
20 years on July 23, 2014

Gina & John Pfau-Moriarty
Spokane, WA
2 years on September 8, 2014

Laura Mentch & Michael Brody
Bozeman, MT
38 years on June 26, 2014

Anne & Jon Williman
Middletown, OH
40 years on August 31, 2014

LOOKING AHEAD

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

Autumn (Current) 2014: Dealing With The Death Of A Loved One With CF.

Winter (February) 2015: Ways To Become A Parent When You Have CF. (Submissions due December 15, 2014.) Are you a parent who has CF? What method did you use to become a parent? Do you have suggestions for others who want to become parents?

Spring (May) 2015: Transitions – Many Types Of Changes. (Submissions due March 15, 2015.) Transitions are all around us. Whether it's moving from pediatric to adult care, leaving home for college or other school, getting married, having children, buying a home, getting a transplant or any other transition, tell us how you handled it.

Summer (August) 2015: What To Expect Post-transplant. (Submissions due June 15, 2015.)

ASK THE ATTORNEY

Answers To Readers' Questions

By Beth Sufian, JD

Many CF *Roundtable* readers have e-mailed and asked for an update on the Arkansas Kalydeco coverage federal civil rights lawsuit. I will provide an update instead of my usual "Ask the Attorney" column.

On June 23, 2014, three girls with cystic fibrosis filed a federal civil rights lawsuit in federal court in Arkansas. The girls had been denied coverage by Arkansas Medicaid in June 2012 for on-label use of Kalydeco. All three girls have G551D, the defective mutation that is corrected by the drug Kalydeco.

This means the girls meet the FDA on-label requirements for coverage of Kalydeco. The FDA has approved Kalydeco for use by individuals who have cystic fibrosis, are over six years of age and have one copy of the mutation G551D. In February 2014 the FDA added eight other gating mutations to the label.

The girls are represented by James Passamano, a partner in the law firm of Sufian & Passamano. The law firm operates the CF Legal Information Hotline and the CF Social Security Project. James became licensed to practice law in Arkansas so that he could represent the three girls. The law firm began representing the girls in April 2014 and is representing the girls for free.

In addition to being my law partner, James Passamano is also my husband. Being married to a person with CF has given James a unique perspective when representing people with CF. When discussing the Arkansas case with others, he has been able to explain how hard it has been to watch me struggle to breathe each and every day of our 26-year marriage. Even opposing counsel in cases he has handled agree that he is a brilliant attorney. He has many offers each year to work at law firms and corporate legal departments, but he chooses to dedicate his legal career to helping people with CF and other medical conditions.

Through the hard work of James, the entire legal team at Sufian & Passamano and the girls' CF Care Center teams, Arkansas Medicaid approved coverage for Kalydeco for two of the girls 30 days after the lawsuit was filed. Prior to the federal lawsuit being filed, the girls had waited two years for a reversal of the June 2012 denial of coverage of Kalydeco by Arkansas Medicaid.

On July 17, 2014, the *Wall Street Journal* published a front page story on the plight of the three girls in Arkansas. It is one of the longest articles published by the *Wall Street Journal* so far this year. There have been shorter versions of the article circulating online. The full article gives an accurate discussion of the issues involved in the case. The article also discusses the failure of Vertex, the company that makes the drug, to provide the drug to the girls while their Medicaid appeals were pending. Read the article at http://online.wsj.com/articles/costly-drug-vertex-is-denied-and-medicaid-patients-sue-1405564205.

BETH SUFIAN

The article featured the story of one of the girls in the lawsuit, a 13-year-old with CF. She told the *Wall Street Journal* reporter, "I feel like they don't care about what's wrong with me, that I'm not as important as everybody else." After two years of waiting for either Arkansas Medicaid or Vertex to provide the drug, it is easy to understand how a 13-year-old or, frankly, any person with CF would feel this way.

Everyone in the United States who has CF and the G551D mutation who wants to take Kalydeco obtained coverage over two years ago from private insurance companies, Medicare and all other state Medicaid agencies. As many research studies have reported, many people with CF who have G551D and take Kalydeco have had positive health benefits.

Without Kalydeco, over the same two-year period, the three girls in Arkansas were each hospitalized numerous times and suffered daily from the effects of CF on their bodies. Readers of this publication know exactly what the girls were experiencing on a daily basis. The difference for the girls in Arkansas was that they knew there was a little blue pill (Kalydeco) that could take away all the suffering.

As a person with CF, I have no idea how they could bear it. I have represented thousands of people with CF who were at the end of their lives.

Of course those cases were upsetting. But those individuals were out of treatment options. Except for the possibility of receiving a lung transplant, there was no pill that could save the lives of those people with CF. The girls in Arkansas are different. There was a pill that could save their lives, but neither Arkansas Medicaid nor Vertex would give them access to the medication.

> *"While the manufacturer's choice of price presents a budget concern for insurers, Arkansas Medicaid has nevertheless now provided the medication to the girls."*

The CF community raised money at fundraisers across the country over many, many years to fund the research that developed Kalydeco. When the price of the drug was announced, $300,000 a year for one person to take the drug, the CF community was afraid the price of the drug would make it difficult for people with CF to obtain access to the treatment. While the manufacturer's choice of price presents a budget concern for insurers, Arkansas Medicaid has nevertheless now provided the medication to the girls.

The CF community knows that there are other companies developing other CFTR potentiators. We will welcome the day the price for drugs like Kalydeco is lower due to competition. Until then, we are thankful Arkansas Medicaid decided not to deprive medically necessary treatment to Arkansans with CF because of the manufacturer's choice of pricing.

As the *Wall Street Journal* article explains, the girls' physicians requested free drug from Vertex starting in 2012. The CF physicians caring for the girls explained that they were working with Arkansas Medicaid to obtain coverage, but in the meantime the girls were sick and in need of Kalydeco. The physicians asked Vertex to provide the drug for free, pending the Medicaid appeals.

As the *Wall Street Journal* reported, "Vertex says it provides free drug to patients without insurance or whose insurance doesn't cover the drug. The company informed the CF Center nurse Medicaid coverage appeals would have to be exhausted before Vertex would provide the drug. But the company later said the assistance program specifically excluded Medicaid patients." (*Wall Street Journal* July 17, 2014, page A10). There is no legal reason to exclude those who have Medicaid from obtaining on-label use of a drug pending an appeal of a denial of coverage. Almost half of all people with CF have Medicaid coverage.

According to the FDA, a drug company can provide on-label use of a drug at its discretion. On July 24, 2014, desperate to obtain Kalydeco for their sick patients, the girls' physicians again requested Vertex provide on-label use of Kalydeco pending the outcome of the legal case.

In July 2014, a group of physicians and scientists asked their colleagues to send letters to Vertex requesting it provide Kalydeco to the girls pending the outcome of their federal lawsuit. Physicians and scientists from around the world sent letters to Vertex asking the company to provide the drug. Well-known blogger Ronnie Sharpe asked the CF community to send letters to Vertex asking it to provide the drug to the girls. Hundreds and hundreds of letters from people with CF, their family members and others who raised the money that funded the Kalydeco research reached the inbox of Vertex's Vice President of Global Affairs, Charles Johnson. The girls, their families and their legal team were encouraged and deeply touched by the outpouring of support from the CF community.

On September 19, 2014, the physician of one of the girls wrote Vertex and again asked Vertex to provide on-label use of the drug to the last girl who was waiting for coverage approval from Arkansas Medicaid. The girl had just had her third hospitalization in 12 months and her second sinus surgery and desperately needed access to Kalydeco. Vertex did not provide the drug to any of the girls. Luckily, we have found good people in Arkansas; people who care about the well-being of their citizens and who understood that the three girls in Arkansas with G551D were suffering and needed immediate access to Kalydeco.

The girls in Arkansas are brave. Each girl cares not only about herself but also about others with cystic fibrosis. Each girl has said that she does not want anyone else to wait two years for life-saving treatment. Each girl wants to advocate for the provision by drug companies of on-label use of CF drugs while appeals of coverage are pending. The federal lawsuit is important to all of us. Any of us could be denied coverage for medical treatment by an insurance company, Medicaid or Medicare. A positive decision in the girls' federal lawsuit will help all of us in our quest for access to the medical treatment we need to fight CF.

Continued on page 11

SPIRIT MEDICINE

A Grieving Spirit

By Isabel Stenzel Byrnes

To stay with that shakiness – to stay with a broken heart, with a rumbling stomach, with the feeling of hopelessness and wanting revenge – that is the path of true awakening. Sticking with that uncertainty, getting the knack of relaxing in the midst of chaos, learning not to panic – this is the spiritual path.
– Pema Chodron

Losing loved ones is part of the human experience. The CF community learns this lesson at a much earlier age than most people in mainstream society. Young people die. We can't say that's not how it's supposed to be, because in our world it happens all the time. We deserve to mourn this injustice.

Each year, on the second night of the CFRI Teen and Adult Retreat, we hold a memorial where we light candles for all our friends who have died. Close friends or family members write the name of a loved one, together with drawings and notes, on an index card for each person who died. We now have more than 250 cards. Many of them are covered in wax, thanks to candle drippings over the years. I once went through the cards and counted the ones I personally knew and interacted with, either at the retreat, former CF camp, or at the hospital. In my 31 years of involvement in the CF community, I've known 123 friends who have died. And, one year ago, I had my most significant loss – my twin sister Ana. It feels like each prior death, whether best friend or cherished acquaintance, has prepared me to face Ana's death.

My task as a "Spirit Medicine" columnist is to express spiritual lessons that may potentially be helpful for those facing cystic fibrosis. So, in this column, I'm very tempted to write a list about how to feel better after the death of a loved one. I am not going to do that; I will leave that article for others to write. I'm seeking to explore the true broken heart of grief.

> *"We survivors have a responsibility to engage in life while carrying around a collective sorrow for our CF community."*

ISABEL STENZEL BYRNES

First, I must be honest and say that living with CF and its surrounding losses is very, very hard. I feel it every day. It brings me down when thoughts of a friend I knew drift into my head. I feel the ache in my chest when I pass an ice cream place I visited with a friend, or when I have to delete another phone number in my contacts.

My losses have left me empty, helpless, befuddled, living in a "Twilight Zone," angry, isolated – all while I continue to engage normally at work, the gym, while traveling, and with friends and family. Grief colors ordinary experiences with a shade of gray whether I want it to or not. It leaves me in a state of confusion; I still look for Ana, expect her call, wonder where she went. I ask myself why I've fought and fought to stay alive – only to be left behind and endure this dark sorrow. I continually feel like I'm being tested – to see how long I can live and how I can find joy in the world without Ana.

I know these feelings are all normal reactions to loss. These "symptoms" (as if it's a disease) will come and go for days, months and years following loss. Grief is the breaking of attachment bonds; it's a primitive biochemical response we have as a result of our evolution as social animals. I have lost a part of myself.

We live in a culture where we need to fix things, to seek pleasure and avoid pain; to value health and youth, and not illness and its wisdom. And yet with grief, there is little we

can do but walk through the muddy waters, get dirty and messy and maybe someday hope we will come out clean. If we avoid or escape grief, it can rear its ugly head somewhere else, physically, mentally or behaviorally. There is no pill we can take; even antidepressants only take an edge off but don't extinguish grief.

Sometimes when I get really overwhelmed, I notice I start to check out of reality. I call this psychic numbing. I start to disbelieve that Ana or so many friends are gone. I wonder if I'm dreaming, I'd rather think of something else, or "I don't care." This reaction is the mind's natural protective mechanism when the emotions are on overload. It's helpful to distance from reality and take in small bits of reminders at a time. This is when I realize I am having grief fatigue and need to immediately partake in life-affirming habits such as listening to music, walking in nature, socializing or doing something creative.

I am aware that grief is a slippery slope towards depression. Depression is the fastest growing fatal disease in the world, and with CF, depression is linked with non-adherence, which is definitely life-shortening. I know there's a list of things-to-do for depression – pets, exercise and storytelling (writing or sharing one's narrative with friends or counselors). But "doing" doesn't always ease the "being" bereaved.

Grief reminds us that we are "spiritual beings having a human experience." Grief is a seed that is planted for spiritual inquiry. I believe that it is in the midst of the emotion of grief where you see the thinnest veil between this world and the other. When someone dies, it is very natural to question, "Where did they go?" It is natural to wonder if they are indeed in a better place, if we'll meet again, or if they visit us in a tangible form like a butterfly or hummingbird. When someone profoundly suffers with CF, it is natural to ask, "Why?" and resist, protest or surrender to this mystery. When the despair, fear and pain of grief drown us, we realize we are not in control, that our egos cannot save us, and that there might be a larger source of comfort. In losing my sister, I've learned what I'm truly made of – an inner flame or will to live that wants to keep burning no matter what has been lost. In my darkest moments, it helps to believe that there's something bigger out there, either orchestrating that "things happen for a reason" or leading me to the source of answers I need to endure. As theologian David Griffin puts it, "God is all-powerful, His power enables people to deal with events beyond their control and He gives us the strength to do those things because He is with us." God sure comes in handy during suffering.

I do not try to fix my grief. It is a life experience that I treasure, because I am still alive to endure this pain and learn from it. Grief becomes the ultimate Life Educator. This crazy roller coaster ride called CF has invited me at a young age to learn universal truths about life: that we are all temporary, that the past, present and future are relative measures of time. Sometimes the weight of grief tries to keep me stuck in the past and immobilized. My loss of Ana and friends has taught me to fully embrace the moment. This is all we have. My past made me who I am today; the people in my past have left permanent impressions. But today I am still me – behind and in front of and through the sadness.

One thing I've learned is to be honest. I need not hide or be ashamed of grief. If tears come, they are just the love in liquid form. Grief is love. I mourn the passing of physical love for Ana and my friends, but my emotional love remains fully alive. We will all die, but our greatest legacy is in our loving. My grief has taught me to be vulnerable and weak so I can receive love, and gain love, so I can then pass it on to others. Grief has taught me to express myself, because being heard, being understood, and being acknowledged is part of the loving.

Lastly, I have a final point to share. My multiple losses have taught me the Golden Rule of Grief: to treat others who are grieving as I'd like to be treated when I'm grieving. My very first "Spirit Medicine" article, in the summer of 2007, was entitled "The Afterlife." It was about my friendship with parents who had lost their CF kids. Thanks to the CF community, I have no lack of mentors – healthy grievers who have the energy to guide me in my grief journey. I remain tremendously thankful to the parents and siblings who have shown me the way; so when my time came to lose Ana, I felt "if others could do it, so could I." These parents taught me the importance of keeping love alive. Jack Lemmon once said, "Death ends a life not a relationship," so staying connected and talking about someone who died helps to maintain a relationship in the present. It's like the dead help us channel love from them back and forth between worlds.

Finally, I grieve for myself and all of you. We survivors have a responsibility to engage in life while carrying around a collective sorrow for our CF community. I feel sorry that we try our best to live optimistically, but then, whether we want it or not, Facebook, the hospital and even this *CF Roundtable* newsletter expose us to blatant news of how others have died and how *we will likely die*. It is difficult and terrifying. Rabbi Harold S. Kushner, in *Conquering Fear: Living Boldly in an Uncertain World*, said, "God's job is not to make sick people

Continued on page 14

SPEEDING PAST 50...

The CF Family In My Heart

By Kathy Russell

Writing about loved ones who have died never is easy. The very act of writing brings back floods of memories. Depending on how long it has been since the person died, the majority of those memories may be either happy or sad. I always hope that the happy will outweigh the sad.

No one in my family, other than myself, has CF. Of course, there may have been ancestors who had it, but I don't know about them. However, I have had many friends who had CF and have died. These people were just like family to me. I can't say that they were like my children, but some of them were as close to me as members of my family. I feel that we were related by love rather than blood.

I first knew that I had CF when I was only 12 years of age. Soon after my diagnosis I experienced the first death of a friend who had CF. It was the cause of many emotions within me. I felt a great sense of loss; I also felt a sense of guilt and I definitely felt relief that I was still alive.

Over the years, as more of the people I knew who had CF died, I began to feel a certain kind numbness about the fact that I also have CF. There were so many people about whom I cared so much that the only way I could deal with it was to ignore the fact that I, too, had CF. Each death was unique to that person and had nothing to do with me. The only connection to me was the love I felt for each of them.

At one point, early in our marriage, Paul and I were going to take a foster child into our home. This little boy (let's call him Dino) had CF and he had very young parents who just couldn't cope with his disease. After he had been in the hospital (where I worked) for more than a year without going home, the docs and social worker were trying to arrange for us to take him. They wanted me to quit my job with the state and just care for Dino. It would save the state money, it would be better for Dino and we thought it would be good for us, too.

Neither Dino nor I had Pseudomonas and we knew each other very well. He always did his therapy better for me than he did for anyone else. The docs and social worker had arranged day trips out for him and we had been a part of those trips. We all got along and had fun together.

There was a lot of paperwork to do to make this happen. The social worker was terrific about keeping things moving. After we had spent months on arrangements and legalities, Dino's parents decided that they would like to try taking him home. I was delighted. I knew that they would be so much happier if they gave it another try.

When he had been home about two months, they brought him back to the hospital in a very ill condition. We doubted that he had much time left. When I admitted him, he was so weak and he had lost a lot of weight. After I weighed him, he didn't want me to put him down on his bed. He clung onto me and just squeezed.

> *"I have had many friends who had CF and have died. These people were just like family to me."*

KATHY RUSSELL

Eventually, I got him settled and left him with his parents. They were able to spend a little while with him before they had to go home. When I went home from work, at 11:30 P.M., he was resting quite comfortably. I knew there was a fair chance that I might never see him again.

The next day when I got to work, the head nurse called me to her office and told me that Dino had died. I was sad but not surprised. His little body had fought the good fight, but he was too ill to go on. Dino was as close to a son as I ever had. I knew him for only about three and a half years. In that time we became very close and he will live in my heart, forever.

Since Dino's death there have been so many other wonderful people I've known who had CF and died much too soon. Until just a few years ago, all of

the people who died were younger than I was at the time. It felt so unfair that such young people had to die. How I wished that each could have lived as long as I have been able to live.

One day, about 15 years ago, I was looking at a paper on which I had written the names of people who had offered to help with *CF Roundtable*. As I scanned the list, I realized that each person on it had died. There were 30 names on that list. I felt as if I had been kicked in the solar plexus. Each of those people had been a friend. All were younger than I and all had been relatively healthy. It made me sad for quite a while. At times life can seem so unfair.

When we started USACFA and began publishing *CF Roundtable*, we all felt fairly healthy. Each of us had issues with our individual cases of CF, but we all managed to live active lives. When Connie Knoles died, that was the first death among the people who started USACFA. Any time I see Jessica Tandy, I think of Connie. We shared a great love and respect for that actress and seeing her reminds me of that shared pleasure. Connie and I were looking forward to my 50th birthday. We were going to celebrate big time. Sadly, she didn't make it that long. I still miss her humor, her talents and her friendship.

Another USACFA death that really hit me hard was that of Joe Kowalski. Joe was like the little brother I always wanted and never had. He and his wife, Doreen, and Paul and I had some great times together. Rarely does a day go by that we don't mention Joe. So many things remind us of him.

Lisa McDonough was another very special person in my life. Many may remember that she started the first newsletter for adults who have CF. It was called *Roundtable* and she produced four issues of it. When she realized that it was too much work for one person, she asked for help. That is how *CF Roundtable* began. The people who called her with offers of help became the founders of USACFA. Lisa went on to write "Spirit Medicine" in this newsletter. She holds a very special place in my heart.

I can't see a hummingbird without thinking of Catharine Martinet. She was a long-time volunteer for USACFA and served as Secretary as well as writer of "Spirit Medicine." I used to tease Cathy's parents that if they ever tired of her, Paul and I would adopt her in a country minute. I feel her around me, often.

Pammie Post is another former Director of USACFA who meant so much to me. Her marvelous sense of humor and her many talents made her an invaluable part of our group. She was so generous with her time and abilities. Paul and I loved getting to see her and being able to call her our friend. Pansies and butterflies make me think of her.

A man who made a deep impression on me was Jack Jacoby, MD. Jack served as Medical Advisor for USACFA. He wrote a wonderful column for this newsletter. In it, he shared some of his insight and caring for others. I always learned from his writing. I felt both honored and pleased to be able to call him my friend. We had wonderful phone calls and shared many laughs. I know that his death was devastating to all the folks with CF who went to him as their doctor at St. Vincent's Hospital Medical Center in New York. There are many little everyday things that remind me of Jack.

There have been so many others who have died. All of them were terrific people. Many of them volunteered to help with producing *CF Roundtable*. These included Theresa who did a lot of the research needed to get USACFA going; Tina who worked on our logo; Ann, Pat, Jeff and Mike who all did what they could to help get the newsletter started. Then there were Ken and Larry who shared editorial duties; Janice who was Secretary; Clay, Susan, another Susan, Jim, Cyndie, another Pat and Dana, all of whom added to the mix of *CF Roundtable* and enriched my life.

There were others who were important to me. Some of those were Ana, Geoff, Nahara, Bracha, Pamela, Marjorie, Tony, Roger, Adina, Barbie, Jim, Barbara and Charlie as well as too many others.

Finally, when I was in my 50s, a friend who was my senior died. Fred was one day older than I. We joked about his seniority and enjoyed discussing our aging. When he died, I felt that I was nearing the "top of the pyramid." Let me explain about that pyramid. I figure that there are more young people who have CF than older people who do. Because of deaths, the numbers get smaller as we get older. Therefore, our ages form a pyramid with fewer people at the top (or older) ages. No one wants to be at the top of that pyramid. It doesn't bode well for survival.

Since Fred's death, Tom, Hal, Kathy and a few others have died. All were older than I. It makes it much harder for me to ignore my mortality. I know it's there and I know that I must put a little gusto into each day.

This has been the toughest article I ever have written. It took me a long time to write it. It brought back so many memories and I needed time to think about it. I miss each of my friends who had CF and have died. My life is better because I knew them. I never will forget them. I wouldn't give up the opportunity to have known these folks. They live in my heart. ▲

Kathy is 70 and has CF. She is Managing Editor of CF Roundtable. *Her contact information is on page 2.*

WELLNESS

Old And Wise

By Julie Desch, MD

Thirty years ago, I began the process of leaving my family behind, as acceptance letters to medical schools far away from my home state of Nebraska began to arrive in my mailbox. Those remaining in NE at the time included my parents and my sister, Kathy, who was dying of cystic fibrosis. My five other siblings were scattered far and wide, but my closest friends remained.

Kathy was the center of my life then, although I don't think she knew it. In her, I saw my future, but somehow was able to keep this fact at a distance and think of it only as her disease and her death. I loved her to pieces, but I was also afraid of her…afraid to get too close and not be able to escape the reality of the horrible disease that I, too, would some day die from.

In the spring of the last year of her life, I visited her in the hospital to discuss the options that were opening up to me. She told me I needed to leave… to go to a medical school where I could do research in CF. To get out of Nebraska and find my own life.

Now I know that she was trying to spare me the sight of her withering body, her rapid decline, the horrible depression that she lived with as her body defied her will at every turn. I decided to go, accepting the offer to Stanford Medical School, and packed to leave for Palo Alto. The lyrics of a popular song back then, "Old and Wise," by Alan Parsons Project, rang in my ears as I left in my Buick Skylark with my husband and dog:

As far as my eyes can see
There are shadows approaching me
And to those I left behind
I wanted you to know
You've always shared my deepest thoughts
You follow where I go

Shadows were approaching me, but at twenty-three years old, I barely acknowledged them. Yet Kathy's were closer…much closer, as I abandoned her to manage them on her own. But I held her with me, and after only a few months of getting used to California, before classes even started, I returned to Nebraska to be with Kathy as she died. At thirty-one years of age, her life was taken. At her funeral, my brother Tom, who also had CF, and I could only hold hands and understand each other in a way that the other siblings couldn't. Our shadows were approaching.

Inexplicably, I made it past thirty-one...way past. I have lived entire lifetimes, as a picture-perfect picket-fence marriage ended, followed by a less than perfect eight-year relationship which resulted in the two best things that have ever occurred, my two sons. Finally, at forty, I found my life partner, and a crazy life of boys, dogs and a two-home relationship ensued.

JULIE DESCH, MD

And then, when I was forty-eight, Tom died. If Kathy was like a mother to me when I was young (she was), then Tom was like a caring, nurturing father. They looked after me as a kid the way my parents couldn't. They understood me. We were the sick ones.

Now, I was the last one standing. The shadows were getting closer. The sadness was overpowering.

And oh when I'm old and wise
Bitter words mean little to me
Autumn winds will blow right through me
And someday in the mist of time
When they asked me if I knew you
I'd smile and say you were a friend of mine
And the sadness would be lifted from my eyes
Oh when I'm old and wise

I don't know when I am considered old and wise. I'm certainly old. In CF years, I'm ancient. I do smile as I think of Kathy and Tom. It is a sad smile, of course. Their lives were stolen. They didn't get three entire lifetimes with kids and dogs and good drugs to keep infections at bay. They didn't live long enough to be guinea pig subjects of new medications that fix the protein that malfunctions in CF. It doesn't seem possible for that sadness to be lifted from my eyes.

As far as my eyes can see
There are shadows surrounding me
And to those I leave behind
I want you all to know
You've always shared my darkest hours

I'll miss you when I go

My body is not quite as functional at age fifty-three as it was thirty years ago when I held Kathy as she took her last breaths. My own shadows are getting closer. Antibiotics don't work as well. Organs are getting tired of toxins. I have been given so much more than I deserve. When I think of Kathy and Tom, I feel so incredibly grateful. First, to have had them as role models and caretakers, but additionally, I have been able to enjoy years and years of family and friends. From old friends from way back in high school to California friends who have been with me from day one of knowing me, caring when I'm sick, helping when they can. I've been able to watch my sons morph into young men, something I never dreamed would happen when I was young. And even though two are gone, I have four remaining siblings who call and worry about me. I have a partner who is willing to live with me to the end…not an easy task, let me assure. I am so fortunate. I don't get it.

And oh, when I'm old and wise
Heavy words that tossed and blew me
Like autumn winds will blow right through me
And someday in the mist of time
When they ask you if you knew me
Remember that you were a friend of mine
As the final curtain falls before my eyes
Oh when I'm old and wise

The final curtain will fall, of course. It has now fallen for both of my parents. I hope that when it happens, in the mist of time, I will finally be able to call myself wise. And I hope you will know that you were all my friends.

As far as my eyes can see ▲

Julie is 53 and is a physician who has CF. You may contact her at: jdesch@usacfa.org.

SUFIAN *continued from page 5*

On October 15, 2014, the attorneys from Sufian & Passamano and their medical experts presented testimony to the Arkansas Drug Utilization Review Board (DUR) in Little Rock, Arkansas. The presentations included information about the medical need for the drug, the legal standard for Medicaid coverage and testimony from the girls. Due to cross-infection concerns, I read the girls' testimony.

Arkansas Medicaid's proposed policy uses the FDA label criteria as its initial coverage criteria for the drug. This is a big victory. However, Arkansas Medicaid's proposed renewal criteria for the drug was concerning as it would have required a person with CF to show that on renewal, after six months of taking the drug, there was stabilization or increase in FEV_1, *AND* a stabilization or increase in weight, *AND* a decrease in exacerbations or hospitalizations. When I presented the girls' testimony I read each girl's plea, "Please do not take Kalydeco away from me." When I presented the testimony of our 14-year-old client, I started to cry. The thought of Kalydeco being taken away from the girls was just too upsetting. While the tears were streaming down my face, I looked up at the DUR Board members and saw that many of them were also in tears. Members of the DUR Board spent a long time questioning one of our medical experts about the renewal criteria. Then the DUR Board voted to remove the *AND*s from the renewal criteria and insert the word *OR*. We are advocating that a last section be added that allows continuation on the drug if there is evidence that it meets a medical need.

On October 16, 2014, the *Wall Street Journal* published a second article about the case and wrote, "Beth Sufian, an attorney representing the Arkansas patients, said she was pleased with the advisory board's recommendation but was concerned the requirements involving prescription renewals, if adopted, could be used to deny the drug. 'Until we see that the renewal criteria is used in a legal way, we feel it is in our clients' best interests to keep the suit alive,' said Ms. Sufian."

Read the entire article at: http://online.wsj.com/articles/advisory-board-backs-requiring-arkansas-to-provide-drug-1413409020d.

When the history of CF is written, what has happened to the girls in Arkansas will be a tragic chapter. We should never again have a time when a 13-year-old with CF thinks that her life is not as important as others'. We should never again have a time when not only a state government fails to provide life-saving medical treatment but also a drug company fails to provide a drug when there is no legal obstacle to doing so and no negative effect on a person's legal claims. We are hopeful that a new chapter will now be written. This chapter will be one of hope for a day when all people with CF have access to the medical treatment needed to fight CF.

Beth is 48 and has CF. She is an attorney who specializes in disability law and she is a Director of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues.

A. Options for Individuals with Excess Assets

An individual with CF may meet the medical eligibility criteria for SSI, but if the individual has over the SSI asset limit of $2,000 for an individual or $3,000 for a couple or family of more than two people, then the individual is over the asset eligibility criteria for SSI.

For example, if a person has $8,000 in the bank he will be over the SSI asset limit. If the individual gives away $6,000 in order to have assets below the $2,000 SSI asset limit, then a transfer penalty is triggered. Giving an asset away for less than its value triggers a transfer penalty causing automatic ineligibility for SSI for a certain period of time – usually up to five years.

If a person wants to spend down his assets to meet the SSI asset eligibility criteria, the money must be spent for items that are used by the SSI applicant. Receipts should be kept in the event Social Security questions the reduction in the assets of the SSI applicant.

The SSI applicant could also create a Special Needs Trust and put the $6,000 in the Special Needs Trust. After the Special Needs Trust has been established and the assets have been deposited in the Special Needs Trust, then the person with CF could apply for SSI. A Special Needs Trust is a specific type of trust created by the Social Security Act. It allows someone with assets over the SSI guidelines to place those assets into a Special Needs Trust. Social Security will not count the assets in the Special Needs Trust when determining eligibility for SSI. No other trust is excluded from being counted by Social Security when determining SSI eligibility. Some attorneys mistakenly believe that as long as assets are in any type of trust the assets will not be counted by Social Security. But only assets in a Special Needs Trust are not counted for purposes of SSI eligibility.

B. Types of Special Needs Trusts

There are three types of Special Needs Trusts.

1.) Self Settled Special Needs Trust (also called First Party Special Needs Trust). This trust contains assets of the individual applying for SSI. The individual applying for SSI must be under age 65 in order to have this type of special needs trust created on his/her behalf. This type of trust is required when an individual puts his/her own assets into the trust in order to qualify for SSI. *See 42 U.S.C. § 1396p(d)(4)(A).*

A First Party Special Needs Trust will terminate only upon the death of the person with CF. Upon the death of the beneficiary, any remaining funds would have to be paid to Medicaid as reimbursement for Medicaid coverage provided to the person during his lifetime. Once Medicaid is reimbursed, any funds left in the Special Needs Trust could be paid to a beneficiary of the trust proceeds.

To establish this type of trust and have the special protection of a Special Needs Trust, the trust must have the following provisions:

The trust must be established with the assets of the beneficiary. The beneficiary would be the individual applying for SSI.

The trust must be established by a parent, grandparent, guardian or court. This means that the SSI applicants cannot set up the trusts themselves.

The beneficiary of the trust must be under age 65 and disabled under Social Security rules.

The trust must be irrevocable, meaning that once it is established, the trust cannot be terminated.

The trust must be administered for the sole benefit of the individual applying for SSI. This means that the trust should only have one beneficiary, and trust funds cannot be used to benefit any other individual.

The trust must contain a provision that upon the death of the beneficiary, all amounts remaining in the trust will be paid to Medicaid to reimburse Medicaid for medical expenses paid during the beneficiary's lifetime. The trust assets cannot pass to a family member of the beneficiary. The assets are paid to Medicaid first to reimburse Medicaid for any funds Medicaid paid for the treatment of the person on SSI. Anything left over could be passed to a family member based on an allocation of the trust funds to designated individuals. *See 42 U.S.C. § 1396p(d)(4)(A).*

2.) Third Party Special Needs Trust. This type of trust does not contain assets of the person applying for SSI. For example, a parent may decide to take his/her own money or assets and put them into a Special Needs Trust for their child. The assets funding the trust did not belong to the child before they went into the trust, therefore, the trust is a third party trust and not a first party trust (discussed above in number 1).

Why does this distinction matter? The distinction matters because third party trusts have less stringent criteria than first party trusts. For example, Third Party Special Needs Trusts DO NOT have to have a provision that Medicaid will be reimbursed upon the death of the beneficiary. Instead any money or assets left in the trust upon the death of the beneficiary can be passed to anyone whom the beneficiary chooses.

3.) Pooled Special Needs Trust. This trust is set up and administered by a nonprofit organization. With this type of Special Needs Trust, upon the death of the trust beneficiary, the assets are retained by the nonprofit agency. *See 42 U.S.C. § 1396p(d)(4)(C).*

C. Use of the Funds to Pay for Beneficiary's "Special Needs"

For the types of Special Needs Trusts discussed above, assets in the trust can be used to pay for the beneficiary's special needs. This could be

almost anything as long as the assets are not used for "basic needs." Food and shelter are considered "basic needs" by the SSA and Medicaid rules. If assets in the trust are used for food or shelter, the amounts spent from the trust are considered "income" to the trust beneficiary and this income could affect eligibility for SSI. However, assets can be used to pay for a variety of things such as education, medical care not covered by Medicaid, home items, home modifications and vehicles.

If trust funds were used in a way that was not for the sole benefit of the person with CF, this could cause the SSA to determine that the Special Needs Trust is no longer excluded for purposes of Medicaid eligibility. If this happened, the person would lose his/her SSI benefit and Medicaid coverage. While this rule is specifically related to a First Party Trust, SSA could also apply the rule to a Third Party trust.

It is very important to talk to an attorney with experience establishing Special Needs Trusts. If the trust is set up and administered properly, the trust can help obtain and maintain SSI coverage.

D. Use of Funds to Pay Family Members

The SSA POMS Section regarding First Party Special Needs Trusts states that trust funds must be used for the sole benefit of the trust beneficiary. This Section specifically cautions that trustee compensation to family members will be scrutinized to ensure that trust funds are being used for the trust beneficiary's sole benefit and not for some other purpose. *See SSA POMS SI 01120.201.*

E. Use of Funds to Pay for Legal or Investment Services

The SSA POMS addresses the use of Special Needs Trust funds for SSI/Medicaid beneficiaries for First Party Trusts but not for Third Party Trusts. *SSA POMS SI 01120.201* of the POMS states that the trust may provide for reasonable costs associated with the investment, legal or other services rendered on behalf of the individual with regard to the trust. See *SSA POMS SI 01120.201 F(2).* This can be helpful for the person with CF who will need to pay costs associated with the Special Needs Trust or other investment services.

F. Examples of SSI/Medicaid Issues Involving Special Needs Trusts

Below are some examples of when a Special Needs Trust could be helpful. These are merely examples and do not guarantee that the example will result in a valid Special Needs Trust being established. There is no guarantee establishing a Special Needs Trust in a similar situation as the one set out in an example will guarantee eligibility for SSI benefits. Please consult with an attorney experienced in Special Needs Trusts to determine the best course of action for your situation.

Example #1: Mike is a 30-year-old man with cystic fibrosis. He is currently receiving SSI and Medicaid benefits. Mike's grandfather passed away and he received an inheritance of $20,000. The receipt of $20,000 will cause Mike to lose SSI and Medicaid because the amount is over the SSI/Medicaid asset limit.

Solution: Mike can have a parent, grandparent, guardian or court set up a FIRST PARTY Special Needs Trust for him. The trust set up for Mike will be a FIRST PARTY trust, because the assets funding the trust ($20,000 inheritance) already belonged to Mike. Mike can then transfer the $20,000 into the Special Needs Trust and keep his SSI and Medicaid benefits. Keep in mind that the receipt of the inheritance makes Mike ineligible for SSI, and during the time that the $20,000 is in Mike's possession, he has assets over the SSI asset limit. Therefore, there may be a loss of SSI and Medicaid benefits until the First Party Trust is established. Mike should have a parent, grandparent, guardian or court set up the First Party Trust as soon as possible.

Example #2: Anna is a disabled child receiving SSI and Medicaid from the state of Texas. Anna's parents want to save money for Anna's college education. However, if the money is put into a standard savings account, it will be considered a countable asset by Medicaid. Once the money saved for college exceeds the Medicaid resource amount, Anna will lose SSI and Medicaid.

Solution: Anna's parents can use their assets to set up a THIRD PARTY Special Needs Trust. The trust will be a THIRD PARTY trust, because the assets funding the trust belonged to Anna's parents. Funds put into the trust will not be counted as a resource by SSI and Medicaid and will allow Anna's parents to save money for Anna's college education.

Example #3: Emma is an 18-year-old female with cystic fibrosis currently receiving SSI and Medicaid. Emma's parents establish a Special Needs Trust for Emma using their assets. The trust is a THIRD PARTY Special Needs Trust. After establishing the trust, Emma's parents have their last will and testament and other non-probate assets (life insurance policies, retirement accounts) list Emma's Special Needs Trust as the beneficiary (instead of naming the beneficiary as Emma).

Solution: By establishing the Special Needs Trust before the parent's death and including a provision in the parent's will and other non-

Continued on page 14

SUFIAN/SOPCHAK *continued from page 13*

probate assets that all inheritances for Emma be put into a Special Needs Trust, there is no loss of SSI and Medicaid benefits after the death of the parent.

Example #4: David is a 16-year-old with cystic fibrosis. David is not receiving SSI or Medicaid benefits. David's parents draft their last will and testament to state that upon their death, if David is receiving SSI or Medicaid benefits, a Special Needs Trust will be created by the assets left for David. Assets that would have gone to David outright will instead fund the newly created Special Needs Trust.

Solution: If at the time of the parent's death, David is receiving SSI or Medicaid, he will not lose these benefits. This is because any assets inherited by David would be placed directly into the Special Needs Trust.

G. Conclusion

Since it is important to protect the SSI benefits of a person with CF, administration of the Special Needs Trust must be handled cautiously in order to prevent a loss of SSI. When a person loses SSI benefits he also loses Medicaid coverage. In 24 states the main way for an adult to obtain Medicaid coverage is to be eligible for SSI benefits. Funds placed into a Special Needs Trust do not count against the SSI income and resource limits provided the trust is **established and administered properly.** The Special Needs Trust can allow a person with CF to qualify for SSI and Medicaid benefits and still keep some assets to help with expenses.

This article only provides information. This article is not meant to be legal advice about a specific situation. No opinion may be inferred or implied in this article. There may be changes in the laws that regulate Special Needs Trusts, so it is important to obtain information from an attorney who understands Special Needs Trust and the unique situation of a person with CF. ▲

Beth is 49 and has CF. She is an attorney who specializes in disability law and she is a Director of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues.

Special contributor Amy Sopchak, JD, is a senior attorney at the law firm of Sufian & Passamano, LLP. Amy has represented thousands of people with CF in Social Security matters. Amy is Assistant Director of the CF Social Security Project, which provides free representation to eligible individuals with CF who are applying for Social Security benefits. She has won all of the cases she has handled for people with CF. The CF community is lucky to have an excellent attorney such as Amy dedicating her legal talent to help those with CF.

BYRNES *continued from page 7*

healthy. That's the doctor's job. God's job is to make sick people brave." I appreciate the charge to have courage. When loved ones die, I can say, "That's their story; I'm living mine." But, truthfully, I *will* have the same fate. I just don't know when… or *exactly* how. Yet, I am reminded that this is the reality of the human predicament for all of us. It's scary, it's uncertain, as no one is immune. Living longer, with CF or not, gives us the potential to mature our minds and spirits, to gain existential intelligence, so that we can find peace with the very source of suffering – fear of death.

Perhaps this article may bring you down. Maybe this topic makes you shed a tear. That's okay. Tears are purifying; let it out. I sure do. As a person with CF, I agree wholeheartedly with Isak Dineson, who said, "The cure for anything is salt water – tears, sweat, or the sea."

Perhaps my sharing my grief makes you fight an urge to reach out and say kind words to make me feel better. But you cannot. Words of consolation often come out as vacuous platitudes. My grief is my own to figure out, just as yours is your own.

The purpose of this "Spirit Medicine" is to show what is. Grief is grief. I am still alive. I am still breathing. I am even happy. But I am grieving. ▲

Isabel Stenzel Byrnes is 42 years old and 10 years post-lung transplant. She lives in Redwood City, CA. Just to share, her TEDx talk can be seen at https://www.youtube.com/watch?v=Dkffpibi-Dc.

Do Not Wipe Your Tears Away
Do not wipe your tears away.
Let them flow down your cheek.
Let them create a stream on your face
To allow the healing waters to flow.
Let the waters cleanse your skin
And wash your face with silk.
Let them caress you lightly
And reveal to you, your heart.
Let the water fall to the earth,
And a tree will grow from it.
Let your tears flow from their depths
And they will release the seeds of your soul.
Do not be careful
Do not be contained
Or proper or polite.
Do not wipe your tears away.
Taste them on your lips
And know that you have graced yourself, today.

—Jodie Senkyrik

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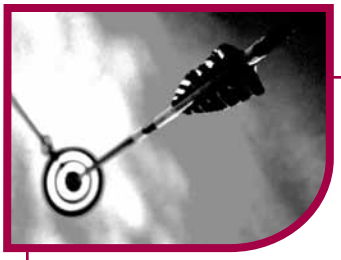
YOU CANNOT FAIL

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

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

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



FOCUS TOPIC

DEALING WITH THE DEATH OF A LOVED ONE WITH CF

Colors Were Brighter

By Mark Levine

I sensed something was wrong from the way our waitress announced to my friend that he had a phone call. It was the night before Thanksgiving in 1993 and a group of us had gathered at a local restaurant, all of us home from school. After taking the call, my friend returned to the table, looked at me, and said, "We have to go." I made a joke about finishing my milk shake. I silently followed my friend to his car. I asked him where we were going and he only answered, "The hospital."

I wanted to ask more but somehow I knew better. The ten-minute drive seemed like an eternity.

My brother had admitted himself to the hospital earlier in the day. He was not feeling well and wanted to get better before he started his next semester in college. At the age of 21, he was three years post bilateral lung transplant and had been experiencing rejection for the previous year.

MARK LEVINE

Before I found David's room, I saw one of his regular nurses crying. She looked at me and simply said, "I'm sorry." When I walked into his room, I saw my parents, our rabbi and one of my mom's good friends, and then I saw my brother lying peacefully on the bed. His battle was over.

It was truly overwhelming. No matter how much you know it may be coming, it is never easy. When I said my goodbyes and leaned over to kiss him on the forehead, he was cold.

I left the hospital that night into a different world, a world without my brother. From here on out, I was an only child. I was about to begin a new chapter in my life, but I was not sure how I was going to handle it.

Thanksgiving has never really been the same. That first Thanksgiving without him was not much of a holiday at all, but we were surrounded by friends and family, which made things easier. I

Information From The Internet...

Compiled by Laura Tillman

NEWS RELEASES

PTC Therapeutics Initiates Confirmatory Phase 3 Clinical Trial of Translarna™ (Ataluren) in Patients with Nonsense Mutation Cystic Fibrosis (nmCF)

PTC Therapeutics, Inc., announced the initiation of a global confirmatory Phase 3 clinical trial of Translarna™ (Ataluren), an investigational new drug, in patients with nonsense mutation cystic fibrosis (nmCF). The Phase 3 confirmatory trial is referred to as ACT CF (Ataluren confirmatory trial in cystic fibrosis) and the primary endpoint is lung function as measured by relative change in percent predicted forced expiratory volume in one second, or FEV_1. Translarna™ (Ataluren) is a protein restoration therapy designed to enable the formation of a functioning protein in patients with genetic disorders caused by a nonsense mutation.
http://tinyurl.com/kmh8ykz

Cystic Fibrosis: additional immune dysfunction discovered

Cystic fibrosis (CF) is a frequent genetic disease affecting the lung and the gastrointestinal tract. Scientists have shown that many of the adult patients with CF in addition lack a cell surface molecule, which is important for immune defense. Cystic fibrosis is due to a mutation of an ion channel which leads to highly viscous mucus and to dysfunction of the lung and the gastrointestinal organs. Since CF

also had music. The songs I listened to that weekend have stayed with me to this day, providing comfort and a level of peace whenever I hear them.

When I finally made it back to graduate school less than two weeks after my brother had passed, everything looked different. Life looked different. Surprisingly, it looked different in a positive way.

As I walked around campus, I noticed things I had never noticed before. Colors were brighter. The air felt crisper. Nature seemed more fascinating. My brother had died, but I was very much alive. I realized that I must appreciate the important things in life, fight the good fight and carry the memory of my brother wherever I go.

This year marks the 21st year since my brother died. He has been gone as long as he was here. It is difficult to believe. On my desk at work, I have a picture of him and me from the summer before he died. I look at it every day. Preserving his memory is as important to me as ever. I talk about him and weave him into conversations when I can. Writing is another way to remember. It is an honor to be able to write about David and keep his memory alive. ▲

> "I realized that I must appreciate the important things in life, fight the good fight and carry the memory of my brother wherever I go."

Mark is 44 and has CF. He is a Director of USACFA and is the Subscription Manager. His contact information is on page 2.

patients frequently suffer from chronic infections, researchers investigated whether these patients might have an additional immune defect. The scientists found that the immunological cell surface molecule HLA-DQ is reduced or absent in many of these patients. Hence, all of the relevant antigen-presenting cells of the immune system are affected.
http://tinyurl.com/l9gj988
http://tinyurl.com/o85plsb
http://tinyurl.com/pazqxvc

Researchers Find Sugar Agent Protects Cells Against CF Superbug Threat Pseudomonas Aeruginosa

A team of research scientists have succeeded in preventing the hospital-acquired bacteria Pseudomonas aeruginosa from entering host cells with the help of a sugar complex. The researchers have identified a sugar complex that binds the bacterial protein LecA. LecA binds the sugar galactose, which is exposed by receptors at the host cell surface. Via LecA, the bacterium attaches itself to the host cell and forces its way in. This allows the bacterium to spread through the body. The researchers developed a molecule that prevents LecA from docking onto the host cell receptors by binding to the bacterial protein with great precision. Binding to host cells via LecA is a significant path of invasion for Pseudomonas aeruginosa in human lung cells. In tests conducted in cell culture, the researchers demonstrated that the germs penetrate into human lung

Continued on page 23

CLUB CF ONLINE

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

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

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

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

A Review Of The 2014 CFRI Educational Conference

By Jeanie Hanley

To be inspired or not to be, to be hopeful or not to be, that is the question. CFRI answered assuredly with hope and inspiration during its 27th Annual CFRI Educational Conference that took place at the beautiful Sofitel hotel in August 2014 in Redwood City, California. The theme was "The Changing Faces of Cystic Fibrosis: Inspiring Hope." Having missed the last several years, due to CF flares and hospitalizations, I was very excited to be able to attend. As always it met every expectation and more. I had remembered the outstanding presentations and speakers of the past, and this conference proved my memory correct. What I had forgotten was the incredible support that abounds from others with and without CF alike and the friendships rekindled and forged.

To give you an inkling of the topics covered: the CFRI conference included eye-opening medical presentations on pain, diabetes, CF metabolic syndrome and newborn screening and risk of cancer. There were also updates on the therapeutic pipeline and the history of CF and what to expect in the future. Fascinating research presentations such as the role of CFTR and novel 3D airway models for CF research were given. Since there was so much great information, I could not possibly include it all here, so please consider investing in the DVDs of these excellent talks, which can be ordered at www.cfri.org.

As if the topics mentioned above weren't enough, adults with CF also graced us with their wisdom. Our very own Isabel Stenzel Byrnes spoke on the importance of adapting to changes in our CF and our resilient characters. Her incredible insights were very welcome, well received and appreciated. Mary Elizabeth Peters, who is a theater artist and CF educator in Boston, also delivered a poignant, poetic and often humorous presentation about pursuing happiness. Anna Modlin was a fabulous Master of Ceremonies for Friday's activities.

JEANIE HANLEY

Saturday night is always extra special with the wine reception and delicious banquet. During the banquet, an exciting raffle took place where a multitude of donated baskets were won by attendees. One, of course, was an awesome "Coffee, Tea and Chocolate" basket donated by USACFA Directors, who each contributed a specialty item from their hometown or state. Our own Julie Desch, MD, was honored with the CFRI Partners in Living Award in Memory of Anabel Stenzel Award. Congratulations, Julie!

The conference final closing on Sunday was not your usual hum-drum closer but instead a rabble-rousing "Ask the Experts: Q & A with Conference Speakers." Members in the audience were provided ample time to have their questions answered by the experts. In this regard the conference was excellent to the very end.

If you have the opportunity to attend the conference in the future, it will be a learning and heartwarming experience. If you are unable to attend for any reason, another option is to watch the live streaming that occurs for a select few of the talks. I guarantee they will deliver a quality presentation whatever the topic. And as previously noted, buying the DVDs would be a great option for those unable to attend.

> "The only thing contagious was the enthusiasm of other attendees."

A note about adhering to infection control guidelines is that cross-infection-control protocols are drilled into all participants' heads and are easy to follow. There is an abundance of masks, gloves, tissues and hand sanitizers that are strewn like candy over tables, in tote bags, baggies, everywhere!

The only thing contagious was the enthusiasm of other attendees. What I did not participate in was CFRI's CF Adult Retreat that occurred the week prior to the conference. After meeting many who did attend, I was infected with the desire to participate next year. I hope to see more of you there at the conference and retreat next year, too. ▲

Jeanie is 52 and has CF. She is a physician who is a Director of USCFA and is the President. Her contact information is on page 2.

PHOTO BY ALEJANDRACHAVERRI

Through My Eyes

Touched by my hands
Nurtured by my words
Comforted by my smile
Encouraged by the sparkle in my eye
My heart has embraced many young lives
The memories are vivid, as is the pain
The paths taken: unique; the journeys:
inspiring
Research rewards us
Hope sustains us

But, in the end, the final destination is always the same.

-K. Shelton, 2002

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

FROM OUR FAMILY PHOTO ALBUM...

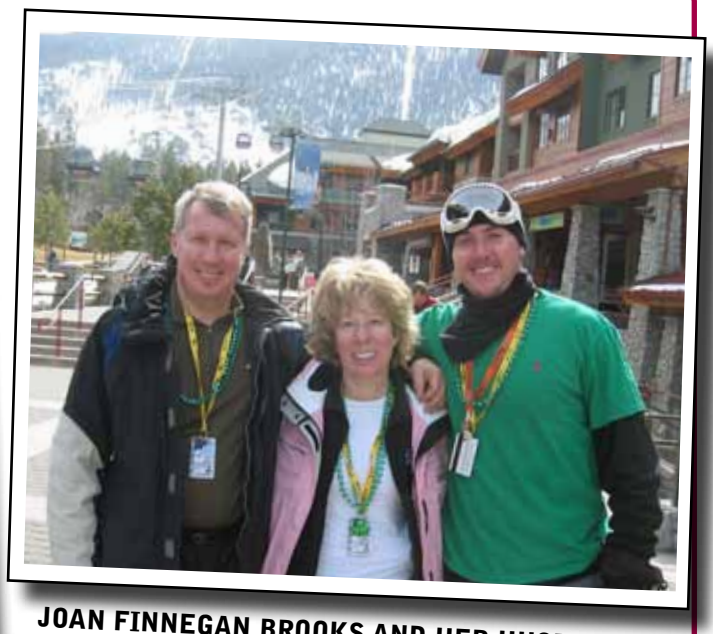
EMILY SCHALLER

JOELLE, BROOKE, ADAM AND MARK LEVINE.

JENNY DOLAN

ISA STENZEL BYRNES AT THE 2014 TRANSPLANT GAMES OF AMERICA, IN HOUSTON, TX.

KAREN VEGA AND HER CHILDREN, LEFT TO RIGHT, LOGAN, KAELI AND JAYDEN. ENJOYING THE MORNING IN MYRTLE BEACH.



JOAN FINNEGAN BROOKS AND HER HUSBAND PETER BROOKS (LEFT) AND THEIR NEPHEW DAN FINNEGAN ON VACATION IN HEAVENLY, LAKE TAHOE.

TOM MARTIN "FLYING" OVER CHAMONIX VALLEY, FRANCE.

JERRY CAHILL

CREATIVE DISENGAGEMENT

Critters In The 'Hood

By Jim Chlebda

When summer cranks up, desert temperatures can be merciless. However, most high desert reptiles seem to get activated by this shift in seasons. The chuckwalla is one of the fascinating creatures I've encountered on morning hikes in 49 Palms Canyon heading to the palm tree oasis. They have to get "warmed up" to around 100 degrees before they start plodding around for their vegetarian dining activities. They love sunning in rocky regions, where they also find security from predators by diving into crevices, puffing their bodies up and out to wedge themselves securely in place 'til danger *(or the intrepid hiker)* has passed. I've shot them in several colors and sizes, from the orange-black one here, to brown with yellow, and grey with green stripes. Whenever I see chuckwallas, I laugh and instantly think: *miniature dinosaurs*.

So far, the zebratail lizard is my favorite. I always cross paths with one or more of these when I hike the park's Scout Trail leading out from Indian Cove heading west. Characteristics include distinct black bands on white tails, beautifully iridescent colors spread across their front legs, back, and sides, intersecting with sharp black belly stripes, and *extremely* long rear toes. They really hoof it when they speed across the hot desert surface to the shade of a near or distant shrub! However, they also seem as curious as I, often stopping to "pose" when I hunker down with my camera for a click or three.

When hiking with visiting friends one weekend on this same trail, I was surprised to see a horned lizard. Later, one of my nephews mentioned that our military has studied them for camouflage tricks to incorporate into their gear—no small wonder, as we all checked out the pics—they have an uncanny knack for blending right in with their sandy home turf!

Got another surprise one afternoon—a pair of desert iguanas raiding a mulberry tree sapling I've had to chickenwire off to keep ever-pesky cottontail rabbits at bay. No problem for these two. While he showed off from below—she climbed up and in, grazing leisurely on tender young leaf shoots. ▲

A CHUCKWALLA ON THE LOOKOUT, 49 PALMS CYN TRAIL.

A ZEBRATAIL LIZARD, ENCOUNTERED ON THE SCOUT TRAIL OUT OF INDIAN COVE.

MASTER OF CAMOUFLAGE, A HORNED LIZARD ON THE SCOUT TRAIL OUT OF INDIAN COVE.

DESERT IGUANAS HELP THEMSELVES TO MULBERRY LEAVES FROM A SAPLING.

Cystic Fibrosis Mothers

Cystic Fibrosis Mothers is a Website dedicated to providing information on parenthood to women with cystic fibrosis around the world. Our aim is to provide a central online resource for the global cystic fibrosis community. It includes personal stories, research articles, advice and links to further sources of information built up over time.

We also provide a private support group on Facebook with more than 500 members worldwide. To visit our Website go to: www.cfmothers.com.

If you would like to join our Facebook support group, please e-mail Karen Vega at: kvega@usacfa.org.

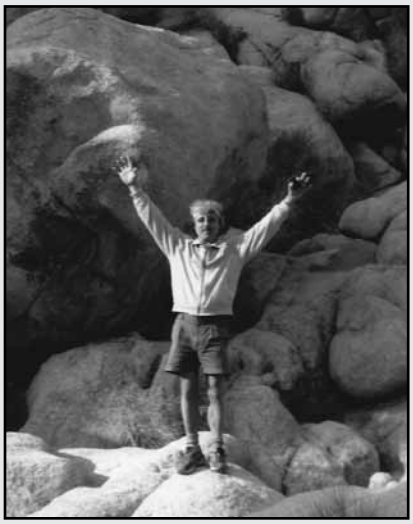
Jim Chlebda
November 26, 1957 –
September 22, 2014

Jim Chlebda was born on November 26, 1957 and died on September 22, 2014. He was 56 years of age. He authored the column "Creative Disengagement," which debuted in the Summer 2013 issue of *CF Roundtable*. In that column he wrote of his life near Joshua Tree National Park in California and included some of his wonderful photography.

Jim told us of how he traded "decades of projects and deadlines in for more time to manage well with CF." He said he always managed to get in a relaxing walk, hike or stroll. He used a digital camera to record the beauty around him. Fortunately for us, he shared some of those photos with the readers of *CF Roundtable*.

He said of himself that he was untethered, relatively unfettered and gainfully unemployed. He edited *Southland Magazine* and created a book publishing firm – Back 40 Publications, which provided a gateway for authors who needed one.

Although his time with *CF Roundtable* was short, we cherish the beauty that he shared with us. We will miss him.



TILLMAN *continued from page 17*

cells 90 percent less often when treated with the sugar-based agent.
http://tinyurl.com/msrk3pd

Sugary Bugs Subvert Antibodies

Pathogenic bacteria are normally destroyed by antibodies, immune proteins that coat the outer surface of the bug, laying a foundation for the deposition of pore-forming "compliment" proteins that poke lethal holes in the bacterial membrane. The antibody response to Pseudomonas can get in its own way. In a subset of infected bronchiectasis patients with particularly poor lung function, an abundance of one specific type of antibody, called IgG2, stripped the blood of its normal bug-killing capacity. The IgG2 proteins bound to extra-long sugars on the bacterial surface, a feature unique to the bugs infecting these patients. When these sugar-specific antibodies were removed, the blood's antibacterial prowess was restored.
http://tinyurl.com/nqvo3wv
http://tinyurl.com/kwgnb4w

Donor-Lung Preservation Device Approved

The XVIVO Perfusion System has been approved by the U.S. Food and Drug Administration to help preserve donor lungs that ultimately may be used for transplant. The device is expected to be used to preserve lungs that initially may not meet transplant criteria but could be used after doctors have been given more time to evaluate the organ. The device can be used to warm donor lungs to the appropriate temperature and flush lung tissue with a sterile solution called STEEN. The device also provides oxygen to the donor organ's cells for up to four hours.
http://tinyurl.com/kgzkanv

New x-ray imaging developed by scientists

Scientists have developed an x-ray imaging system that enables researchers to see "live" how effective treatments are for cystic fibrosis. The new x-ray imaging method allows researchers to look at soft tissue structures like the lungs. The new imaging method allows scientists for the first time to non-invasively see how the treatment is working "live" on the airway surface.
http://www.healthcanal.com/

Agile Sciences lands $1.5M NIH grant for cystic fibrosis therapy

Agile Sciences has landed a $1.5 million National Institutes of Health grant to help develop its cystic fibrosis therapy derived from sea sponges. The company's technology platform capitalizes on a chemical produced by the sea sponge to break up bacterial clumps called biofilms. The latest NIH grant supports preclinical development of Agile's proprietary Agilyte anti-biofilm molecules for treating lung infections of cystic fibrosis patients.
http://tinyurl.com/l4ezr2k

Pitt study: Cystic fibrosis could be two diseases

People with unexplained pancreatic problems might have a previously unknown type of cystic fibrosis that doesn't affect the lungs, according to researchers at the University of Pittsburgh. This second kind of cystic fibrosis is caused by nine major mutations that had gone unnoticed by the medical community.
http://tinyurl.com/lv5zyjb
http://tinyurl.com/po2kmpk

New Study Links Cystic Fibrosis with a High Percentage of Patients Infected with Mold

This study found that almost 50% of adult patients who suffer from cystic fibrosis (CF) are also infected with Aspergillus, a common type of fungi found in the environment and in water-damaged buildings. The new

Continued on page 25

PROTECTING WHAT MATTERS

Being Prepared For The Death Of A Loved One

By Mark Manginelli

Benjamin Franklin once said, "In this world nothing can be said to be certain, except death and taxes." Unfortunately, all of us will most likely have to endure the loss of a loved one throughout our lives, often on numerous occasions. How we effectively deal with such a loss is paramount in our recovery process and, to some extent, foreshadows how we will live the rest of our own lives without them. While we all cope with loss in different ways emotionally, there are important financial components that people often overlook that could help make things more manageable. Death never comes at a convenient time, but being able to identify the difficult conversations and decisions with the luxury of time allows us to carefully consider these things without the unneeded pressure of crisis or catastrophe.

The focus of this article is to educate our CF community and shine light on the importance of implementing a plan *before* that fateful day comes. In my personal and professional experiences, most people do not enjoy talking about estate and life insurance planning; there aren't many people who enjoy creating a hypothetical scenario without their loved ones in the storyline. I've found that once the conversations are initiated and plans are formulated, every party involved comes out of it with a renewed sense of relief and peace of mind. On the contrary, we can all think of friends, family members, colleagues etc. who didn't plan properly and wished they could turn back the hands of time. Unfortunately, I've witnessed personal friends in my local CF community lose their battle to CF without any plan at all. The legal and financial aftermath that their family had to endure was heartbreaking. There are many imperative factors to consider, especially when cystic fibrosis is a part of the equation.

Whether you are a caregiver for an individual with CF or have CF yourself, the first (and sometimes the hardest) thing to do is to start the conversation with all necessary parties. There will always be something else going on in our busy lives that could hinder these conversations or put them on the back burner but, unfortunately, major life events can happen without warning. By investing a few hours, you could save yourself from significant future anguish during an already difficult time. Understanding where to start and what to know with regard to planning out the financial future of your loved ones is often something that most people struggle with. With that said, I have created a basic and general checklist to help kick-start your planning process. This checklist should be revisited annually because important details change when you get married or divorced, have children, move, change jobs etc. It is also important to keep in mind that every plan is unique in its own way and may be more, or less, complicated.

> *"While we all cope with loss in different ways emotionally, there are important financial components that people often overlook that could help make things more manageable."*

MARK MANGINELLI

Draft a Will

- Appoint a Durable Power of Attorney – This person will handle specific health, legal and financial responsibilities.
- Guardian – Who will take care of your minor children in the event of your death?
- Distribution of assets and personal items.
- Trusts – Protect government benefits and/or ensure monies are distributed exactly as you wish.

Draft a Living Will (Advance health care directive)

- Health care power of attorney – who will make any necessary decisions for you if you are incapacitated and unable to do so?
- Written instructions about type of funeral or memorial service you desire.
- Considerations on where and how you will be treated if you become terminally ill.

Obtain Life Insurance

- Caregivers for people with CF should consider life insurance in the event they predecease their dependent. How will their dependent continue to receive the same care without the presence of the caregiver?
- Individuals with CF – This topic is more complicated because most life insurance companies do not cover individuals with cystic fibrosis. There are, however, a few life insurance companies out there that do offer policies to individuals with CF on a contingent basis. Because of the wide spectrum of CF instances, everyone would have to inquire about their individual eligibility.

Money – Are your financials organized and prepared for unexpected events?

- How will income be affected with the death or disability of a caregiver or CF dependent alike?
- Does your employer offer short- or long-term disability and have you enrolled in it?
- Have you set up an "emergency fund" or have a safety net to cover expenses for an extended period of time?
- What long-term savings plans have you already set up or need to implement?

Details – How will your loved ones organize their new life without you?

- Organize your important details
- Passwords
- Official documents – Birth certificates, marriage certificate, social security numbers etc.
- Financial Info – bank/investment accounts, home/auto insurance, mortgage, credit cards etc.

Although this list is a mere overview of the things we should consider while planning for our family's future, the complexity and scope of a plan may be more or less in depth than what this checklist offers. It is important to seek the advice from trusted legal and financial resources to help successfully implement these items. I hope that when our respective day comes and the grieving process of losing a loved one with CF begins, it doesn't bring extra suffering because we were not prepared. A wise man once said, "Most people don't plan to fail; they fail to plan."▲

Mark is 28 and has CF. He lives in Edison, NJ. You may contact him at:mmanginelli@usacfa.org.

TILLMAN *continued from page 23*

study brings to light the importance of CF patients avoiding exposure to elevated levels of Aspergillus in their homes, schools and businesses. Aspergillus is also frequently found in household dust and on building materials in properties that have suffered moisture damage and mold growth.
http://tinyurl.com/ojd7t68

In CF, two drugs are no better than one

Researchers explain why efforts to improve CF treatment by combining ivacaftor with new drugs have failed. Ivacaftor increases mutant CFTR's activity, but it only works when CFTR is on the cell surface. The new drugs under development bring mutant CFTR to the surface, but combining the two types of drugs has not been effective because ivacaftor also makes CFTR less stable, so cells remove it quickly from their membranes.
http://tinyurl.com/pu5oquu
http://tinyurl.com/nhpzsde
http://tinyurl.com/kf9p5xw
http://www.hcplive.com/

Quadruple Dosing of Doripenem Safe in Patients with CF

Quadrupling the US Food and Drug Administration's approved dose of the antibiotic doripenem in patients with cystic fibrosis (CF) and acute respiratory infection is safe and could offer a new therapeutic option for those with advanced disease and evidence of resistant bacteria in their lungs.
http://tinyurl.com/lof5wru

Medical startup seeks FDA approval for lung-disease product

The product is called Syntopical System I and is a device for purifying the air, water and surfaces in the home. Asthma & Allergy and its partners are seeking FDA approval for a "humanitarian device exemption," which would expedite approval. The Syntopical product already has received the FDA's "humanitarian use" designation, a first step in the expedited process.
http://tinyurl.com/kuolrjt

Continued on page 30

PARENTING

The Greatest Reward In The World

NEW COLUMN

By Karen Vega

Hello, readers. My name is Karen, and I'd like to introduce you to a new column discussing starting a family and parenting when you have cystic fibrosis. I am very excited about this column as I am a mother of three young children and love sharing my stories to motivate and support others. The column will feature stories from many guest writers, both men and women, covering a variety of topics about fertility, pregnancy, adoption, surrogacy and unique challenges that we as CF patients face while being parents. To introduce myself, I'd like to start by telling my journey to motherhood.

From the time I was a little girl, I always knew I wanted to be a mom. So when my husband and I got married, deciding to start a family was an easy decision. We did, however, put a lot of planning into starting a family. Luckily my lung function was stable (with FEV_1 in the mid-high 70%s) and I am pancreatic sufficient. My husband had the genetic sequencing done to rule out being a carrier of the CF gene. I didn't feel I could successfully manage my health, children and a career, so we decided that when the time came I would quit my job and focus on my health and my kids.

I became pregnant about six months after we started trying and got the surprise of a lifetime at my first ultrasound appointment. I learned that I was carrying twins! My OB and CF doctors were both very nervous since a multiple pregnancy definitely puts more stress on one's body and lungs, but they had confidence that I would have a successful pregnancy.

Despite being classified as a high risk, the pregnancy was textbook and uncomplicated. I was able to use my Vest throughout the pregnancy, loosening and eventually unbuckling the bottom clips, and I continued nebulizing HyperSal and Pulmozyme. For the most part my lungs remained stable, apart from a drop at the end due to the lack of space in my lungs and inability to clear them properly. I had one course of IVs at 29 weeks, after developing a productive cough, but managed to continue to work from home until my boys were born.

With no warning I went into preterm labor just shy of 32 weeks. I went to the hospital, and, 48 hours after my contractions had started, the doctor told me that I needed an emergency C-section because one of my sons was showing signs of distress. I was wheeled into the OR and quickly both boys were born. The doctors made sure they were stable, and I got to briefly see them as they were rushed to the NICU.

Jayden was born at 4 pounds 10 ounces and was on Bi-PAP for just a few hours. He had no preemie complications other than being jaundiced and having to learn how to eat properly (suck, swallow, breathe). He was in the NICU for four weeks. Logan (the twin who was showing distress) was born at 4 pounds and had a few more problems. He was on a ventilator for a few days and Bi-PAP after that. He had a PDA (hole in the heart), needed a blood transfusion and also had jaundice. On Christmas Eve, at 11 days old, the boys got to reunite in the same bed for the first time since birth. Jayden was in the NICU for six weeks and his PDA closed on its own by the time he was two months old.

KAREN VEGA

For me, coughing after my emergency section was rough. I decided to go on IVs after I delivered to get rid of any infection and be in better shape for when I got home. I was also able to stay in-patient an extra week so I could be with the boys while they were in NICU. It took me about three weeks to feel normal again after the surgery.

The year following the twins' birth was an extremely rough time for me. On top of caring for newborn twins, I was breastfeeding, had kidney stones for the first time and was diagnosed with the flu when they were seven months old. I wasn't sleeping well for a long time. Fitting in two treatments a day was near impossible. July 2009, when I had the flu, my lung function was the worst I've ever seen it, 35% below my baseline.

I just didn't have time to think about anything other than the babies at that point in time. I had very little energy to do anything, and, looking back I was probably somewhat depressed feeling trapped inside my

home that first winter. It was extremely difficult to go anywhere alone with two babies. It was almost impossible to go even food shopping with two infants in car seats. I was very lucky to have my mom with me to help while my husband was at work.

After another exacerbation and two weeks of IVs in December 2009, something clicked inside me and I knew I needed to be more pro-active. My lungs weren't magically going to get better on their own without some hard work on my part. For the first time I was actually scared for my future. I was angry at myself for not trying harder, for underestimating what CF can do in such a short amount of time. I want to live, for myself, for my husband, for my children. And not just live, I want to have energy, run with them, dance with them, play with them. I woke up one day and joined a fitness center and began swimming again. For the first time in probably close to seven years I was exercising.

I decided to give myself the biggest push I ever have and signed up to run the New York City Marathon for Team Boomer later that year. On the day I signed up, I couldn't run a mile without stopping because I was out of breath. But on November 7, 2010, I completed 19 miles of the marathon before being forced to stop due to cold and exhaustion.

Maybe if I hadn't had a chest infection at the time I would have completed it, but from the beginning of this journey it was never about crossing the finish line. It was about challenging myself and pushing myself to be a better me. And I succeeded. I ran 19 miles when just seven months earlier I couldn't even run a mile. I was able to increase my lung function by over 50% from that low I had 18 months earlier. I learned so much about myself and I became a stronger person.

When I got pregnant with my daughter in 2011, I was in much better physical shape than my prior pregnancy. I didn't need any IVs and felt great the entire pregnancy. I stayed very active with my toddlers, and I also swam and did prenatal yoga. I had a repeat C-section at 40 weeks and recovered in two days. I was out doing normal activities as soon as I left the hospital. I attribute this to being in much better physical shape and continuing to exercise my entire pregnancy.

My daughter is currently two years old and my twins are 5½ years old. Becoming a mother has had a positive influence on my health. Although it becomes harder to fit in treatments and nebulizers, I have three of the most important reasons to keep fighting. I don't beat myself up over missing a treatment every now and then, I do my best to work them into my schedule, and, overall we have a good routine going. Our new family activity is bike riding, so we all get to work out and have fun together.

Becoming a parent isn't just about getting your body in good shape to become pregnant. It is a commitment and a lifestyle change and promise to yourself and your children that you will do everything possible not only for them but for yourself so that you can enjoy every moment. It means having a sick plan and support in place for when you aren't feeling well.

But all that planning and hard work has the greatest reward in the world – hearing your children say, "I love you, Mommy." ▲

Karen is 33 and is a Director of USACFA. Her contact information is on page 2.

Pay It Forward

Again, our readers continue to amaze us. Our first fundraising campaign continues on – beyond our expectations! We are overwhelmed by your generosity and support of our efforts to make *CF Roundtable* available for free. We thank you for your continuing endorsement of our endeavor.

Below is a list of those who continue to "pay it forward":

Norma Van Auken
(In memory of Nahara Mau)
Rick Birkner
Anne Bodlee
(In memory of Nahara Mau)
Sandy Defenbaugh
(In memory of Nahara Mau)
Adam Levy
Norma Ludwig
Elzie & Martha Malone
(In memory of Glenn Alan and Jamie Faye)
Jan Meek
Richard & Dorothy Millikan
(In memory of Nahara Mau)
Barbara Williams (In memory of Nahara Mau)

IN THE SPOTLIGHT
With Joan Finnegan Brooks

By Andrea Eisenman and Jeanie Hanley

If you are a long-time reader of *CF Roundtable*, you may recall that Joan Finnegan Brooks is a former director of USACFA. She has been, and remains to this day, very involved with the CF community. She cut her teeth at an early age as a volunteer with the Cystic Fibrosis Foundation (CFF) as one of three siblings with cystic fibrosis (CF) and incredibly concerned and involved parents. They were among early volunteers birthing the crusade of finding a cure for CF at the creation of the CFF more than 50 years ago. With all of her experience with the Foundation, an excellent education in economics and work in marketing and financial services, Joan started her own consulting business. Along with working, and being married to a wonderful husband, Joan keeps to a strict treatment schedule so as to not miss any medications. As well as taking her meds, she believes exercise is key to staying healthy with CF. When you meet Joan in person, you can tell she exercises. Although it is mainly for her health it is also part of staying positive with CF. And an added bonus is, she looks fabulous. Meet our newest star. Spotlight, please!

JOAN FINNEGAN BROOKS

Age: 54

Diagnosed: at 1 month of age after I developed pneumonia.

As an infant with CF, explain how you were adopted, and ultimately saved.

At the time of my adoption, my parents had already suffered the heartbreak of losing their two-and-a-half-year-old daughter, Peggy, to cystic fibrosis. They had another child, John, who also had CF and was six years old at the time I joined my family. Only my parents' oldest son, Pat, didn't have CF and was 12 years old when I came into the picture.

My mom got a call from another CF parent and a long-time family friend – Doris Tulcin, who ended up being the Chairman of the Board of Trustees of CFF many years later – asking my mom if she could call the foster parent who was caring for me at the time. My mother hung up the phone and told my father that there was a little girl with cystic fibrosis in foster care. My father declared, "that little girl needs us!" and three weeks later, I was adopted into a loving, wonderful family. I feel so lucky and blessed to have joined my extraordinary parents and brothers!

How did your parents get involved with the CFF?

My parents were among early volunteers who started the CFF in 1955. Other parents of children with CF banded together to try to find doctors to treat their children, fund research etc. This was the very early and dark days in CF history. These parents were desperate to help their children. Children didn't live very long, and my parents had already lost a daughter to CF in 1952.

I've always been involved with the CFF, for as long as I can remember. It was almost as if I had been born into the Foundation. My family always participated in fundraisers and would do anything that the doctors thought would be helpful to advance CF science, like donate blood to look for the gene. Remember, this is back in the 1960s and 1970s before a lot was known about CF science. As a young adult moving from New York to Boston, one of the first things I did was contact the local CFF chapter to get involved. I felt "right at home" and joined their board of directors. Having CF is challenging, but the gift of community with so many wonderful people is the "silver lining." Some of my fondest memories are associated with great times I've had at various CFF events. It's always been like an extended family to me.

Has your faith helped you?

My faith has definitely helped me through life's challenges, CF-related and otherwise. I find my faith a great source of comfort, strength and renewal.

What do you do for a living and how did you decide on this work?

After I graduated from Brown University with a degree in economics, I began a career in the financial services industry. I had a successful 17-year career that encompassed great variety – securities trading, marketing, investment products etc. During

the same time, I was always active with the CFF and loved the science and story behind the success of the CFF. I really wanted to combine my business skills with my passion, which led me to found my own consulting practice, Patient-Focused Market Research. I consult with and provide key insights into drug development and marketing programs for companies working on treatments for CF. It's so important for companies to integrate patient, caregiver and clinician perspectives in their programs.

What are pros and cons of working for oneself?

Because I work for myself, I have a great deal of flexibility to control my schedule, which is really helpful to get all my CF treatments done. I do a lot of my work in collaboration with other people and I really enjoy that aspect of my work – it keeps me from feeling too isolated. The downside of working for yourself is on days that you don't feel well, no one else is there to do the work. On balance, I enjoy the freedom of my schedule and the interest in my work.

When were you diagnosed with diabetes? How is it controlled?

I was diagnosed with CF-related diabetes (CFRD) in 1987. I was blessed to have a wonderful friend who had Type-1 diabetes and she really helped me learn about and manage diabetes. I've taken insulin injections since my diagnosis and it has helped me tremendously with my energy level and lung function. While no one likes to have another thing to deal with in addition to CF, learning to live with and control diabetes is worth a better future. I started using an insulin pump about seven years ago and it's given me a great deal of flexibility, since I rarely eat or exercise at the same time each day. I also learned how to estimate carbohydrates in my meals so I can take the right amount of insulin. I always find that the diabetes nurse educator is a great resource to help with the "day-to-day" adjustments you may need to make.

How did you meet your husband?

My friend with Type-1 diabetes introduced me to my husband, Peter. They worked together at IBM and we met at one of her parties. Peter and I started playing racquetball first before we started dating – he never let me win a game!

How long have you been married?

Peter and I will celebrate our 25th anniversary in 2014!

What do you do for exercise?

I typically run and cycle with a great group of friends who make exercise a nice social activity. I'm not really a winter runner so I'll go to the YMCA to use the cardio machines. I'm also trying to get back to weight training and core strength exercises.

Have you always been so physically active?

I've been active since I began high school. Back in my days growing up, physical activity wasn't really viewed as necessary or helpful for people with CF. But I loved playing field hockey and noticed that on game and practice days, my lungs felt much better and I was able to clear secretions much easier. From that experience, I have always done some high-intensity aerobic exercise. Through the years, I've tried many things: racquetball, running, cardio kickboxing, aerobics, Zumba, Pilates, yoga, cycling etc.

Are you excited about the new therapies for treating CF?

I am very hopeful and optimistic that new therapies targeting the CFTR protein will help me. I am very supportive of the CFF's drug discovery efforts and thrilled to see Kalydeco help people with the G551D mutation. I am always looking to participate in clinical trials and hope everyone else considers joining so that we can get new therapies as soon as possible!

Do you take antibiotics on a regular basis?

I take some oral antibiotics on a regular basis and do IVs when I need to. I try to finish up my IVs at home if possible.

What do you do for fun?

I enjoy my time with my husband, family and friends. Recently, Peter and I went with a group of friends on a riverboat cruise in Germany to celebrate a friend's 60th birthday! It was fantastic and a great way to travel for me, since I only had to unpack all my medical "stuff" once! I also like being at home, reading a book or playing the piano.

Discuss how you have been able to stay healthy enough to benefit from recent medical discoveries, such as Pulmozyme and hypertonic saline:

I am so lucky to benefit from those therapies to maintain my health. People don't appreciate what it was like without those drugs – I'm old enough to remember the dark days before we discovered the CF gene. It was bleak! My routine of exercising has really helped me maintain my lung function over the years so that when new treatments are developed, I can benefit from them. That's been the case for me with hypertonic saline and Pulmozyme.

What do you mean by "setting the clock back" in terms of non-adherence?

I know how difficult it is to maintain our CF treatments, but every time we skip them, it's the same as "turning the clock back" in time to the dark days of CF history. I lost my brother John to CF when he was 15 years old in 1969, before we had any drugs specifically developed for CF. Even though we have to do a lot to

Continued on page 30

BROOKS *continued from page 29*

maintain our health, it's a privilege to have drugs and treatments that actually help us!

Do you still volunteer?

It's important for me to stay involved with the CF community, from whom I've received so much! I serve on various committees with the CFF and am always ready to be helpful to other people, families, companies and clinicians dealing with CF. Locally, I stay involved (by phone and e-mail) with my local CFF chapter.

How do you find the time to do all of your treatments, work and be married?

Like everyone else, it can be a daily struggle to keep up with CF treatments, exercise, eating well, resting and family responsibilities. My best days are when I follow a schedule of sorts, so that I'm not leaving my treatments to chance "whenever I have the time," which can be never! I'm lucky to have a wonderful support network and husband. So on days when I'm overwhelmed or not feeling great, I can get some help.

What are your goals?

As simplistic as it sounds, my goal is just to be healthy enough to do the things I still enjoy. I aspire to have a nice word for people, an inviting home, good friends and close family and fun adventures!

What helps you stay motivated and compliant?

Thinking about my future keeps me motivated to keep up with my treatments. I look forward to seeing my nephew, cousins and friends' kids grow up and want to enjoy being with Peter, family and friends. I am also motivated by the solemn vow I made to myself many years ago: I would never look back on my life and wish that I had taken better care of myself. The only way I know to fulfill that promise is to focus on my actions in the present, and do all my treatments today, tomorrow and the next day.

Where do you see yourself in the next five years?

Ideally, I'd like to travel a bit more, but most importantly, be able to celebrate my 30th anniversary with my husband! I'd be quite content to continue on the path I'm on now in terms of work, exercise, family and friends.

Anything inspiring to add?

As my Mom used to remind me all the time, make health your top priority in your life, for without it, all your other choices are limited. I do what I have to do to stay healthy so that I can do what I want to do with my life. Choose to be optimistic about your health: consider yourself a healthy person with periods of sickness, instead of a sick person with periods of health. Remembering that sick times are not the "norm" of my life motivates me to return to the "healthy" version of myself by doing what is necessary to optimize my lung function and weight.

Joan can be contacted by e-mail or phone at: joanfbrooks@p-fmr.com, 617-312-0931. ▲

Jeanie Hanley is 51 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2. Andrea Eisenman is 49 and has CF. She is a Director of USACFA and is the Executive Editor of CF Roundtable *and Webmaster. Her contact information is on page 2.*

If you would like to be interviewed for In The Spotlight, please contact either Andrea or Jeanie.

TILLMAN *continued from page 25*

Trinity scientists discover how to attack a bacteria that causes cystic fibrosis infections

Pseudomonas aeruginosa is a resilient and adaptable species of bacteria that causes disease by infecting damaged tissue. The bacteria is known as an "opportunistic pathogen," which is the common cause of lung infections in cystic fibrosis patients. The bacteria produces a moist, viscous, slimy "bio-film" that is hard to attack. Alginate, a major component of this bio-film, is made in the bacterial cells and is passed out via a pore in the outer membrane. This outer membrane helps to "ring-fence" each bacterium from its external environment, while the pores in it provide controlled "gateways" through which the alginate can exit. The research looked at the hole that is punched through the membrane – which is just a few atoms wide – which could let antibiotics penetrate the bacteria. If the movement can be blocked and the growth of bacteria can be slowed down, it would be a welcome development.

http://tinyurl.com/m6gb2hw

Exercise Capacity of CF Patients Focus of Three Projects at Georgia Regents University

Sildenafil, antioxidants and gene therapy for cystic fibrosis treatment are part of an arsenal of studies. Research has shown that cardiac function may negatively affect exercise capacity of cystic fibrosis patients even if patients have adequate lung function. Although sildenafil and antioxidants are not being pursued as sustainable treatment options at present, they are tools to understand the reasons for poor exercise capacity. The research group is administering low doses of sildenafil to adult patients multiple times daily for four weeks. Before and after treatment, patients are tested for endothelial func-

CONVERSATION CORNER

Anatomy Of A CF Table

By Jenny Dolan

JENNY DOLAN'S CF TABLE.

When I first moved to Los Angeles, my other half and I lived in a 500-square-foot apartment. My CF paraphernalia would be strewn all over the house – neb cups on the counter, compressor machine shoved behind the couch, and where did I put that pesky inhaler again?

When we moved into our next apartment, I vowed to make a place in my home for CF. I promised myself that I wouldn't hide my CF equipment when company came over, either. Hence my CF Table!

I keep my Flutter and inhaler in a small bowl that my oldest friend, Evonne, gave me.

I haven't lived near family in five years – my parents live in Chicago. I display a photo of my mom and me, to remind myself that I'm not alone in this.

My best friend, Paige, gave me the little pink lungs. Paige was recently diagnosed with multiple sclerosis. In the card, she said we were like characters in the *Wizard of Oz*, me without healthy lungs, and her without a healthy brain, skipping down the yellow brick road. Paige has more grace than anyone I know, and the lungs remind me of her.

I think it's important to make room for CF in our lives – both literally and metaphorically. Creating a CF Table, and keeping it stocked with meaningful objects, makes my treatments more enjoyable. Not surprisingly, I find that I'm more compliant with the treatments now, too.

How do you make room for CF in your home? I'd love to hear from you. Shoot me an e-mail at: JennyLDolan@gmail.com, or visit my blog at www.thecontentmentchronicles.com. ▲

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tion and exercise capacity. It is thought sildenafil will increase blood flow and retrain the blood vessels to respond more normally to exercise. Another strategy for studying exercise capacity in cystic fibrosis patients is administering a combination of over-the-counter antioxidants. Patients with high levels of oxidative stress and chronic inflammation are the target audience for therapy. Also of interest to the group is gene therapy to directly address dysfunctional chloride channels that contribute to the sticky, glue-like mucus that accumulates in the lungs. The team is collecting baseline measures of endothelial function and exercise capacity for patients who qualify for combination gene therapy that is projected to be available next year.

http://tinyurl.com/mjr83gr
http://tinyurl.com/ox5eovo
http://tinyurl.com/mk52hpv
http://tinyurl.com/n9fjvj6

TREATMENTS

Ibuprofen rescues mutant cystic fibrosis transmembrane conductance regulator trafficking. Graeme W. Carlile, Renaud Robert, Julie Goepp, Elizabeth Matthes, Jie Liao, Bart Kus, Sean D. Macknight, Daniela Rotin,

Continued on page 37

Jerry & Em's Great CF Bike Adventure: He Said/She Said

Welcome to Jerry and Em's Great CF Bike Adventure where Jerry Cahill and Emily Schaller will ride their bikes 500 miles to promote awareness about cystic fibrosis (CF) and the importance of exercise for those who have CF. They will start in Cary, NC, at the Chiesi USA headquarters and end at Johns Hopkins Hospital in Baltimore, MD, while making stops in between.

We will find out the rationale behind the story, what their goals are for the CF community and themselves. They will share how important exercise has been in their lives and how they first started.

Both Jerry and Em have CF and are very involved in the CF community. They need very little introduction. But in case you have been living under a rock for the last 10 years, briefly: Jerry Cahill has been a volunteer with Boomer Esiason Foundation (BEF) for the last 10 years. He has started many programs at BEF to benefit those with CF and highlights with podcasts issues we face every day (www.jerrycahill.com). Also with grants from BEF, he helps people receive scholarships to go to college and assists people with transplant grants to help financially with incidental expenses. He is post-transplant two years and is 58 years young. Emily, 32, has been playing drums in a band called Hellen. She plays at fundraisers for CF, raises awareness and funds to further CF research. She also started a non-profit organization called Rock CF that began with using music to raise money but now includes walks, runs and biking to encourage healthy exercise for everyone.

Bike To Breathe: Jerry & Em's Great CF Bike Adventure is one of the first events for BEF using biking to illustrate the importance of fitness for those with CF and their families.

JERRY CAHILL AND EMILY SCHALLER.

What brought you two together to bike 500 miles?

Jerry: Boomer, CEO of BEF, is always supportive about the importance of exercise so I was writing grants to pharmaceutical companies to promote such programs. And I had been speaking to Em about possibly "teaming up" on an athletic event. Running has become more difficult for my knees, I have been re-inventing my exercise routine with biking and I thought, let's do a bike adventure to raise awareness about exercise. Em said yes and the process started several months ago and here we are… BOOM!

Em: Jerry did. He said, "Em, let's plan a bike trip" and I said, "Okay." It was that easy. Next thing I know I've committed to riding 500 miles!

What do you hope to accomplish at your stops along the way?

Jerry: Raise awareness about CF and the success of lung transplantation, inspire others and challenge ourselves. I believe that we ALL need to challenge ourselves. "You Cannot Fail" and "We Are All Heroes Of Our Own Story." For me, it's about the journey, about sportsmanship, about friendships made along the way.

Em: The stops will probably be my favorite part, not only because it will be a break from riding, but it will allow us to connect with people. We have plans to speak at a few of our stops but it's the natural organic connections that will come up with the townspeople and others who are wondering what the heck we are doing.

What are your goals for Bike to Breathe?

Jerry: Get people moving! RE-INVENT the world of CF and post-transplant with exercise! When I grew up, exercise NEVER was encouraged for people with CF.

Em: I definitely hope that we can shed even more light on the benefits of exercise for everyone, especially those with CF. That being said, we are not telling people with CF to go bike 500 miles or run a marathon. Just set some type of goal that is attainable and go for it!

How did you prepare for this ride?

Jerry: A lot of biking, cross-training and nutrition…and getting assistance from my awesome team of doctors at New York Presbyterian Hospital, a part of Columbia University Medical Center (CUMC).

I always exercise, but for this "adventure" I was more focused on biking and training for months. With biking long rides it's all about "saddle time" – spending hours on the bike. At first it's not about speed – just get on and "spin."

Em: Riding. Riding. Riding, and more riding. I've done a few 300-mile bike trips before, but adding a few more days and miles on with this ride is going to be interesting. At least I'll have Jerry and a great crew to make it entertaining.

What have you learned during your training?

Jerry: I learned a lot about nutrition and proper hydration and carbs before and during the long rides. Although I thought I was mentally tough, this training has made me more so. RELENTLESS.

Em: When training for any endurance activity, nutrition and hydration are key. I always find that I have to re-tune my eating and drinking on the bike to make sure I'm dialed in and won't bonk-out on the road.

How has exercise changed your life? When did you start and why?

Jerry: Exercise is the single most important thing in my life that has kept me healthy. All I can say is that I am still here at 58, two years post-transplant, and I believe it's all about the power of exercise, my positive family and my great doctors.

I started to exercise when I was diagnosed with CF at around age 10, even though my doctors didn't recommend it. My parents were uncertain about how long I would live; at that time life expectancy for people with CF was 16. They wanted me to spend time with my brothers doing sports. Little did they know it is what has extended my life.

Em: Exercise has become the single most important thing that I have done to extend and improve my life. I used to be a couch potato. Actually, no, I was the couch that the potato sat on. Since beginning to run and cycle in 2007, my FEV_1 has increased drastically, my hospital stays were greatly reduced and I felt better than ever. I got to the point where I was sick of being sick, sick of the hospital stays, sick of feeling tired all the time and sick of not knowing how my future would end up. When I first started I couldn't even run one block. Thankfully I stuck with it and now value every step, pedal stroke, downward dog or pull-up – which I have yet to accomplish.

What advice would you give people to get them to exercise?

Jerry: Just get out there and start moving. It's not about running a marathon or biking 500 miles, just doing something physical makes people feel better. Coughing during exercise is good – "Clear the airways!"

Em: If you are tired of being tired, sick, moody or whatever, YOU can make the change and be the one to improve these feelings. Start slow, but start. Find something that you like to do and do it. The benefit far outweighs the alternatives.

Who or what inspires you to stay as active as you are?

Jerry: Staying healthy inspires me. I hate to be sick so I am very driven to stay fit and it's fun. Also the high school track athletes I coach keep me on my toes.

Em: Jerry does. He is the one person who inspired me to start exercising and keep going.

You are both avid runners, why bike?

Jerry: I have always liked biking and did a half-ironman relay where I did the biking part and I enjoyed it. Also, since I am getting older and running is harder on my knees after pole vaulting for over 35 years, two NYC marathons, and four half marathons, it was time for me to RE-INVENT myself again. Life, especially with CF, is all about re-inventing yourself.

Em: I actually started running and cycling in 2007. So both are equally awesome! Biking long distances takes a little less of a toll on the body and we are getting old.

How do you reinvent yourself and why?

Jerry: Life changes all the time so if you do not go with change and you let obstacles (like CF) stop you, you are stuck and cannot move forward. I don't like obstacles in my life and if they are there, and they are, I figure out a way to go around them – RE-INVENT!

Em: My theory is that we all should be the best person that we can be. I didn't know what my potential was until I really dug in and felt the difference in my health and outlook on life after starting to exercise. The old me was pretty awesome but very unhealthy. I'm so glad that I went through this "life-style change," seven years ago, instead of a mid-life crisis.

What would you like to add, if anything?

Jerry: I like to live by the Fighter's Creed as much as possible and it pertains to EVERYONE no matter where you are in life.

"Life is a fight round by round. I have the choice to fight or quit. I am responsible for the direction of my dreams. I will work hard and I will keep going no matter what. From this day forward I choose to fight."

– The Fighter's Creed ▲

New Airway Clearance Therapies: MetaNeb® And AerobiKa®

By Laura Mentch

My growing cache of meds and equipment for CF care includes these for airway clearance: a flutter valve, two Acapella devices – one green and one blue – and a vest. None has hit the mark for me. Unless I am having an exacerbation, I bring up little sputum during treatments. I've recently been able to use new airway clearance therapies that are more effective for me, MetaNeb and AerobiKa. I hope that you will have a chance to see if they make a difference for you.

The MetaNeb® System

About a year and a half ago I was introduced to MetaNeb, a new therapy from Hill-Rom, while in the hospital. Of all of the therapies I've used in the hospital – vest, percussor and IPV – the MetaNeb works best for me. I started by using it during the mid-day treatments with a nebulizer and hypertonic saline (HTS), but have been able to use it during all four of my daily in-patient treatments. The mouthpiece (or mask) with nebulizer is attached to the MetaNeb machine that is connected to the oxygen source. While the vibration with the IPV was helpful, the two-phase action of the MetaNeb has been the most effective treatment I've used.

Treatment with MetaNeb takes 10 minutes with two repeating phases: the first, CPEP (continuous positive expiratory pressure), expands the lungs and distributes the medication. The second phase during exhalation provides resistance and internal vibration CHFO, (continuous high frequency oscillation) to encourage mucus clearance. Exhalation with the MetaNeb takes effort and I can feel its action in my lungs. My typically scant collection of sputum increased markedly with the MetaNeb. I wanted one for home! Alas, it requires an oxygen source, which I do not have.

AerobiKa® Oscillating Positive Expiratory Pressure (OPEP) Therapy System

Several months ago I read a Facebook post from another patient at my clinic excited about a new device our respiratory therapist had given her, AerobiKa. I asked about it at my next visit and went home with my own. As with the MetaNeb, I feel this treatment vibrates my lungs from the inside and helps me bring up more sputum than with other therapies. It is easy to use, the size of your palm and portable.

AerobiKa can be used alone or with a nebulizer and medication. I use it twice daily with nebulized HTS before the vest. Either AerobiKa or vest could be used alone, but I like combining the two approaches. With inhalation the HTS is drawn into my lungs, when I exhale the resistance and vibration loosens and moves mucus. Huff coughing, a third technique, helps bring it up. AerobiKa cleaning and care is the same as for other nebs. I am grateful to have my AerobiKa in the front of my CF shelf while I remain hopeful that the MetaNeb may, in time, become practical for use at home.

http://www.hill-rom.com/usa/Products/Category/Respiratory-Care/MetaNeb-System

http://www.monaghanmed.com/products/consumer/aerobika-oscillating-positive-expiratory-pressure-opep-therapy-system ▲

LAURA MENTCH USING HER AEROBIKA WHILE INHALING HYPERTONIC SALINE.

Laura is 61 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Cystic Fibrosis Adult Stem-Cell Treatment

By Angelo DiStefano

What a concept: I woke up because I was finished sleeping. Let me explain, but a little background is in order first: One afternoon in March of 2011, I sat in the clinic with my cystic fibrosis doctor at the Cystic Fibrosis Clinic in Nashville. My health was going downhill fast. Even after three back-to-back IV antibiotic home-infusion treatments, my lung function continued to drop. My IV regimens usually run for two weeks at a time. On September 15, 2010, my FEV_1 was 27 percent. The medical community generally accepts FEV_1 as the primary indicator of lung function for cystic fibrosis patients.

At the end of December 2010, I had the first of the three IV treatments due to an exacerbation. On January 12, 2011, my FEV_1 was 26 percent. So the doctors did another IV treatment. At the end of the two-week treatment, I was still not improving, so they continued right into the third round of IV antibiotics. Even though I was not improving, we had to stop the IV medications because the drugs were beyond a toxic level in my body. By March 16, 2011, my FEV_1 had dropped another two points to 24 percent.

In the meanwhile, I had been researching adult stems for nearly two years. My research produced information about stem-cell treatments in other parts of the world, but up to this point, I found nothing for cystic fibrosis and nothing in the United States. I was not totally averse to leaving the country for a treatment, but I wanted a United States connection. In February, I learned about a cardio-vascular surgeon in Florida who was doing adult stem-cell treatments in the Dominican Republic. After talking with his office in early March, I learned that he had recently treated a cystic fibrosis patient, who's breathing starting improving only three weeks after the treatment.

ANGELO DISTEFANO AND WIFE, GARI, WITH DR. GREKOS. ANGELO IS HOLDING THE STEM CELLS FROM BONE MARROW THAT HAD BEEN ACTIVATED WITH GROWTH FACTORS.

In March, the CF doctor put me on a new inhaled antibiotic, Cayston. This drug almost killed me and caused my lung function to drop even further. My body was sliding into a slow death, and I did not even have enough lung function to speak in complete sentences without stopping to catch my breath. I was gasping for air even at rest.

I made a decision with my wife. We would proceed forward with the stem cells. Through family and business associates, my wife raised the money we needed for the treatment, some ancillary expenses, and our living costs for a few months – more than $60,000 in total. I knew that I had only months, if not weeks, to live, and we were praying that I would live long enough to get the treatment.

My last CF Clinic visit before the stem-cell treatment was in April. My FEV_1 had dropped to 18 percent. I was reluctant to say anything to the cystic fibrosis doctor about stem cells, but I had to say something. I wanted to talk about it with my doctor, but these doctors had not been amenable to discussing alternative treatments in the past.

I asked my doctor what she knew about adult stem-cell treatments. I was not prepared for the response.

She said, "They don't work."

So I asked, "How do you know?"

She became visibly upset and reiterated, "They absolutely do not work."

She did not answer my question. Thinking she had some evidence, I asked, "Are there any clinical trials for stems cells with cystic fibrosis?"

Now she was having trouble maintaining her composure. She said, "No. Stem cells do not work!" and she actually turned her chair to turn her back to me and continued typing on the keyboard. She refused to talk any further. After she finished typing she just left the room.

I had my first adult stem-cell transplant on May 3 and 4, 2011. The

Continued on page 36

process involved taking bone marrow from me and my mother. In the case of cystic fibrosis, a genetically matched donor is necessary because my stem cells will produce new lung tissue that is still affected by cystic fibrosis. I am not sure exactly of the process, but somehow by mixing the cells from the donor with my own, my body sees the cells as my own and does not reject them. Immediate family is necessary for a close-enough genetic match.

I almost died several times before and just after the treatment. Not from the treatment, but from being at end-stage cystic fibrosis. You can read more about that in the archives of my personal blog www.angelodistefano.com. I got worse before I got better, but about five or six weeks after the treatment, I began to feel my breathing function improve. The improvement was gradual, but continued for several months.

By December 7, 2011, my FEV_1 was back up to 25 percent. The other measurements of my pulmonary functions tests improved even more dramatically. After about a year, the stem cells quit producing new tissue, and I needed a follow-up treatment. That was early June of 2012. I should have gone for the third treatment in May or June of 2013, but money was an issue. I started losing ground because of the progressive nature of cystic fibrosis.

On July 31, 2014, I received my third treatment. This third treatment included a new procedure, adding growth factors to the stem cells. As of this writing (about three weeks post treatment), I am just beginning to feel my breathing function start to improve. Because of the new procedure, I expect even greater results this time.

Before Stem-Cell Treatment

Most noticeable was the improvement in my overall well-being. Before the treatment, I was struggling to breathe at rest. I had trouble sleeping due to breathing issues and severe headaches. I basically had to keep a high level of ibuprofen in my bloodstream to combat the headaches. I was too weak to eat any more, and my brain was in a constant fog. I was no longer getting out of the house because just showering or walking across the living room would almost make me pass out. My wife had to help me bathe.

Eating became a real issue. Because of cystic fibrosis and the related malabsorption, I need a huge amount of nutrition. But within a few minutes of eating, the energy taken by the digestion process caused me to become weak, almost to the point of passing out. And my chest was so tight and filled with congestion, that just a small amount of food would press on my lungs and make it even more difficult to breathe. So I was not even getting enough nutrition for my body to maintain life. I was sleeping sixteen to eighteen hours a day, although it was not really sleep. After the few hours that I actually slept, I would awaken more exhausted than before going to bed.

After Stem-Cell Treatment

At the peak of improvement, after the second treatment, I was sleeping well and able to function on only eight hours of sleep. I was getting out of the house and able to drive myself, because my brain clarity had returned. I was eating well and gaining weight. I was able to meet people at events all day and into the evening, and do so without a portable oxygen tank. I was no longer tethered to an oxygen hose twenty-four hours a day. I was able to dance again with my wife, if only for one song at a time. The stem cells reversed osteoporosis and strengthened my immune system. In three years since the first treatment, I caught a cold only once in the first year, and got over it in three days without any exacerbations. I have not had any IV antibiotic treatments since the spring of 2011 because I have not needed them.

We moved to south Florida in the summer of 2011 to be closer to my stem-cell doctor and to get away from the Nashville team who ran out of options for me and actually told my wife that she would just have to deal with the fact that I would be dead soon. I now have another cystic fibrosis doctor in south Florida who is great to work with. He still thinks there is no proof that the stem cells work, but he cannot explain why I have not needed a tune-up (IV antibiotics) in three years. We had a more comprehensive set of pulmonary functions tests run just before this third treatment to establish my baseline numbers. We will do the same tests at the three-month mark and the six-month mark from July 31. My cystic fibrosis doctor, who says I have no proof, is helping to develop the proof to his satisfaction.

Back to the Explanation

My second stem-cell treatment was in June 2012 and I should have had the third in the spring of 2013. I was not able to get the third treatment until July 31, 2014, and by then I had lost a lot of ground. My FEV_1 has dropped to 20%. I am struggling again and I get out of breath easily, although breathing is not difficult like it was in 2011. I don't get out of the house much and need to sleep about 15 hours a day. So I get to bed about eight or nine and wake up about noon. The headaches are back, so that interferes with sleep, but I am nowhere near as bad as in 2011, before the first treatment. My immune system is still strong, and my bone density has improved—reversed during the last three years from osteoporosis to osteopenia. But probably the most insidious problem of late is getting the intense headaches during sleep.

The headaches started coming back

in early 2014, but I was able to control them with ibuprofen or acetaminophen. I believe the headaches are caused by a number of contributors, but a major source is severe hypoxia – low oxygen to the brain – and muscle spasms also caused by low oxygenation. In June, the headaches got out of control.

This became a typical night:

I would start my night by taking two acetaminophen tablets at eight or nine when I went to bed. By midnight, a headache woke me and I took one tablet. At two, a more intense headache woke me and I took another tablet, sat up and massaged my neck and shoulders for ten minutes before lying down. I awoke again at four to an even more intense headache, so I took two tablets. This time I was pressing against my temples and massaging pressure points in my neck and shoulders, trying to squeeze and break up the spasms. I also tried to breathe deeply to oxygenate my body. Twenty minutes later, I lay down. The next one came at five thirty and was even more agonizing. I was lying down again at six. And awake again by seven-thirty – the headache was so excruciating, that I did not want to lie down again, so I got out of bed and dragged myself to the kitchen for a cup of coffee. I was no longer able to sleep until noon to get my 15 hours of sleep. And I was getting almost no sleep during the night, so instead of being restored during the night, I was getting more run down. I was more exhausted in the morning than at night before bedtime. I came to dread going to bed, knowing what was to come.

So imagine my relief two nights ago when I awoke at midnight, maybe out of habit, but NO HEADACHE. I took one tablet just to be safe. Then I did not awaken again until about six A.M. I had only a slight headache – very slight, so I took another tablet and felt my neck and shoulders. They were soft, not in spasms. Back to sleep, yeah! And then I awoke at ten yesterday morning. Not because of a headache, but just because I was finished sleeping. Wow, what a concept! Praise the Lord!

Last night I was trying to figure out what was different. With all the medications and supplements I take, when something gets out of balance, I use the process of elimination to determine the culprit. But nothing had changed. Then after dinner last night, it hit me. The previous night I had a mini cannoli for dessert and had to do some reading and editing. So what goes well with both of those? A shot of espresso! Did you know that coffee is a natural vasodilator. Increased blood flow means better oxygenation and voila! It works better than acetaminophen to relieve headaches. I tried it again last night, and slept much better. So my remedy for headaches is an evening shot of espresso for the Sicilian. Kind of poetic, don't you think?

Now that I can sleep, I look forward to continued improvement in the coming weeks as the stem cells do their work. As my breathing improves, my body oxygenation will also improve, and the headaches will no longer be a problem. The same thing happened after the first stem-cell treatment. I will post updates as appropriate. I am writing a book about my journey, so look for news in the months ahead. My goal is to raise awareness about adult stem-cell therapy for cystic fibrosis and generate funding to provide financial assistance to patients who need the treatments.

If you want to follow along for updates and news about adult stem-cell treatments, or if you have questions about the adult stem-cell treatments, you can follow and reach me on Facebook at: https://www.facebook.com/angelo.distefano2, Facebook Group: https://www.facebook.com/groups/Distefano/ or my personal blog: www.AngeloDiStefano.com. ▲

TILLMAN *continued from page 31*

John W. Hanrahan, David Y. Thomas. Journal of Cystic Fibrosis. Published Online: June 25, 2014

Small molecules as shown by VX809 can rescue the mislocalization of F508del-CFTR. The aim of this study was to identify correctors with a clinical history and their targets of action. Ibuprofen was identified as a CFTR corrector. These studies show that ibuprofen is a CFTR corrector and that it causes correction by COX-1 inhibition. Hence ibuprofen may be suitable to be part of a future CF combination therapy.
http://tinyurl.com/mvqc456

Ceftazidime-avibactam: an evidence-based review of its pharmacology and potential use in the treatment of Gram-negative bacterial infections. Lagacé-Wiens P, Walkty A, Karlowsky JA. Core Evidence, 08/11/2014

In this review, the authors summarize the in-vitro data, pharmacology, mechanisms of action and resistance, and clinical trial data relating to the use of this agent combined with ceftazidime for the treatment of Gram-negative bacterial infections. The addition of avibactam to ceftazidime improves its in-vitro activity against Enterobacteriaceae and Pseudomonas aeruginosa.
http://tinyurl.com/ng4675d

Omalizumab: a new treatment option for allergic bronchopulmonary asper-

Continued on page 38

gillosis in patients with cystic fibrosis. Lehmann S, Pfannenstiel C, Friedrichs F, Kröger K, Wagner N, Tenbrock K. Ther Adv Respir Dis. 2014 Aug 21.

Allergic bronchopulmonary aspergillosis (ABPA) is a severe complication in patients with cystic fibrosis (CF), resulting in deterioration of lung function and impairment of overall prognosis. Standard therapy consists of high dosage, long-term corticosteroid treatment. This carries the risk of serious side effects such as immune suppression, diabetes and osteoporosis. Antifungal drugs such as itraconazole may cause interactions with other drugs and drug levels need to be monitored. Omalizumab treatment has been tried in several case studies. Omalizumab has the potential to be an additional and solitary treatment option in patients with CF and ABPA. Early onset treatment may be beneficial and patients with early stage of lung disease seem to benefit more.
http://tinyurl.com/n8zrkse

Inhaled versus nebulized tobramycin: A real world comparison in adult cystic fibrosis (CF). Harrison MJ, McCarthy M, Fleming C, Hickey C, Shortt C, Eustace JA, Murphy DM, Plant BJ. Journal Cystic Fibrosis. 2014 May 9.

There are no published data on real-life clinical experience comparing inhaled antibiotic therapy via new rapid delivery systems with nebulised antibiotic therapy in CF. This real-world study compares safety, effectiveness and tolerability using tobramycin inhaled powder (TIP) versus tobramycin inhaled solution (TIS). Inhaled powder tobramycin in CF is associated with improved adherence, tolerability and decreased exacerbation rates compared to nebulised treatment in real-life practice.
http://tinyurl.com/p8qfjgl

Antibiotic developed 50 years ago may be the key to fighting "superbugs." University at Buffalo Health and Medicine News, 06/19/2014. University at Buffalo. Science Daily, 23 June 2014.

Scientists at the University at Buffalo are turning to an old class of antibiotics to fight new superbugs resistant to modern medicine. Developed more than 50 years ago, polymyxins were not subject to modern antibiotic drug development standards. And they have proved to be toxic to both the kidneys and nervous system. But they're also effective against superbugs such as Acinetobacter baumannii, Pseudomonas aeruginosa, Klebsiella pneumoniae and other gram-negative bacteria that are resistant to all current antibiotics. The aim of the project is to evaluate novel dosing regimens for polymyxin combinations to maximize antibacterial activity and to minimize the emergence of resistance and toxicity.
http://tinyurl.com/msynr8t

ASPERGILLUS

Aspergillus sensitization or carriage in cystic fibrosis patients. Fillaux J, et al. The Pediatric Infectious Disease Journal, 06/16/2014

The authors aimed to identify possible predictive factors for patients who become sensitized to Aspergillus fumigatus (Af), compared with a control group of non-sensitized Af carriers. Patients with non-severe mutation but low FEV_1 baselines are becoming colonized with Af or patients with high FEV_1 baselines who present with severe mutation are more susceptible to the Af sensitization and then to the presentation of an allergic bronchopulmonary aspergillosis event.
http://tinyurl.com/o3uwqvo

LUNG TRANSPLANT

Impact of pulmonary hypertension on survival in patients with cystic fibrosis undergoing lung transplantation: An analysis of the UNOS registry. Don Hayes Jr., Robert S. Higgins, Stephen Kirkby, Karen S. McCoy, Allison M. Wehr, Amy M. Lehman, Bryan A. Whitson. Published Online: January 02, 2014

A high prevalence of pulmonary hypertension was found in CF patients prior to lung transplantation. The researchers found that there is no strong evidence suggesting that it significantly alters the risk of death in CF patients after LTx.
http://tinyurl.com/nrez53g ▲

Laura is 66 and has CF. She is a former Director of USACFA. She and her husband, Lew, live in Northville, MI.

In Memory

James Richard Chlebda, 56
Joshua Tree, CA
Died on September 22, 2014

Geoffrey David Earnest, 38
Portland, OR
Died on May 8, 2014

Gerry Roy, 48
Tavernier, FL
Died on May 9, 2014

Jeffrey Tews, 60
Estero, FL
Died on June 28, 2014

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

We enjoy reading *CF Roundtable*. My husband was elected coroner. This money was left in his fund, so we are sending it to you in memory of our children Glenn & Jamie Faye. It is a comfort to know things are better now for CF Children.

Elzie & Martha Malone
Red Bay, AL

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- Please notify us immediately of any address changes. Returned mail wastes money and delays mailings.
- We would like to act as a referral source for active adult support groups. Please send us your group name, leader's name and phone number, number and age range of your members and geographical area covered, and we will add you to our referral list.
- Please let us know of the major occurrences in your life (*e.g.*, *marriages*, *births*, *completion of educational degrees or training*, *career advancement*, *transplants*, *anniversaries*, *birthdays*) and we will print your information in **Milestones**.
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- You can reach **USACFA** and ***CF Roundtable*** at anytime by e-mail at **cfroundtable@usacfa.org**
- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq., 712 Main, Suite 2130, Houston, Texas 77005. E-mail: CFLegal@sufianpassamano.com.**
- You may subscribe at www.cfroundtable.com

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

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

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

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

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