CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS SUMMER 2010

Prednisone – Don't Leave Home Without It!

By Michelle Thornell

My husband and I have always dreamed of travel. When our children were very young we took them camping all over Alberta and British Columbia. When they were a little older, we took them to San Diego one spring and Prince Edward Island the next. We were going to continue on in this manner but, unfortunately, cystic fibrosis (CF) reared its ugly head and my health began a rapid decline. We were content to keep our travels close to home and continued to go camping every summer and skiing every winter. As the children grew and my husband took more and more trips for business, I longed to go with him.

We were nervous about traveling with CF. All the stuff you had to take, what if you got sick while you were away and the fact that travel insurance would not cover a pre-existing condition such as CF, were all reasons we gave for staying close to home.

When my health declined to the point where I had to retire from my job and go on disability, I figured that I would have to table any big travel dreams. A life-threatening episode actually made me turn around and promise myself that I would see more of the world. Our first trip after this was a safe, all-inclusive trip to Cancun, Mexico. It was a lot of fun and we did every excursion we could. The bug had bitten us and we began to make other plans.

Our first real trip, in our eyes, was a trip to Costa Rica in 2005 for our 25th wedding anniversary. We decided that we would book only our first night's hotel and just wing it from there. We flew into San Jose and rented a car and had the best vacation ever. With no deadlines to meet, we were free to travel at our own speed. If I needed a rest day, we took one. We fell in love with the rain forest and indulged in our passion of bird watching. My fascination of snakes and bugs was indulged to the fullest. We snorkeled and took boat trips and had some lovely beach days. The two weeks flew by and we couldn't wait to plan another vacation.

In 2007 our daughter took some time off from University and traveled to Thailand for a month. Helping her plan her trip planted the seeds of desire and so that spring, off we went. Because my husband travels a lot, he is able to accumulate Aeroplan miles and so we were able to make this journey in business class, very helpful for someone with health issues! We traveled with backpacks and, again, made no reservations. The Buddhist religion was truly fascinating to explore and I found it very peaceful and uplifting. We visited many temples and I was blessed by a monk. We rode on the backs of elephants, with me feeling sorry for the elephant every step of

Continued on page 21

INSIDE THIS ISSUE

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United States Adult Cystic Fibrosis Assn., Inc.
PO Box 1618
Gresham, OR 97030-0519
Voice mail or FAX (503) 669-3561
E-mail: cfroundtable@usacfa.org
www.cfroundtable.com

EDITOR'S NOTES

Summer has arrived with a vengeance. We have just had one of the hottest Julys in memory. So, enjoy the beaches, camping and summer vacations. Also to enjoy are the summer foods of picnicking, barbequing and cool drinks. Remember to stay hydrated and use sun-block during these sunny days.

We have some more changes on our Board of Directors. **Rich DeNagel** and **Julie Desch** chose not to run for re-election. However, Julie will continue to write her column, *Wellness* and Rich will do at least one more *Unplugged* column. We thank both of them for their participation. We are happy to welcome **Anne Williman** and **Jen Eisenmann** as new Directors on our Board. I know they will be positive additions to our group. You can read more about them on page 29.

In this issue, our Focus topic is: "Becoming a Parent with CF." **Jeanie Hanley** starts us off with her article, Sweet Music. She discusses her concerns of the effect of her CF on her children's social lives while wanting to also be part of family-time as she does her treatments. **Steve Gonzalez** weighs in with his feelings that having a child while having CF is complicated yet incredibly rewarding. Both **Bracha Witonsky** and **A. Nonymous** tell their stories about how thrilled they are to be mothers while balancing their CF and the care of their kids. In her article, Parenting Isn't for Sissies, **Anne Williman** writes about her and her husband's experience of private adoption and raising three kids while maintaining her energy and health.

On our cover, **Michelle Thornell** writes about her many travel experiences and how being well-prepared while on a trip can lead to a successful vacation. **Janice White** writes in *Voices from the Roundtable* of becoming a Hero of Hope and all she does to stay busy and healthy.

On to our columnists: **Beth Sufian** detangles and explains the new healthcare reform laws in *Ask The Attorney*. She tells how these provisions will positively affect the CF population and when we can expect these changes to take effect. Most of us have experienced a loss of someone we knew with CF and wondered, "why them?" **Isa Stenzel Bynres** ponders, Why Bad Things happen to Good People with CF, in *Spirit Medicine*. *In Speeding Past 50*, **Kathy Russell** includes two parent's writings regarding their hopes and fears for their children's adult lives. **Julie Desch** writes humorously about having children, from "The Decision" to the "Teenage Years", in her *Wellness* column. Following that, in *A Deep Breath In*, Debbie Ajini writes about being a parent to oneself and knowing when it is time to retire from the workforce. And finally, in *Unplugged*, **Rich Denagel** interviews a 21-year-old Japanese man with CF who has many hurdles to overcome just to maintain his health in a country where CF is a rarity. It leaves one with many things to appreciate. In *Information from The Internet*, **Laura Tillman** lists discoveries from new and upcoming CF medications to reproduction in people with CF. Something to inspire hope in anyone.

– Andrea Eisenman

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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

Paul Feld
Florissant, MO
53 on May 9, 2010

Steven Gonzalez
Rockville Centre, NY
45 on November 22, 2009

Wedding

Paul & Kristi Feld
Florissant, MO
19 years on June 1, 2010

Steven & Bridget Gonzalez
Rockville Centre, NY
16 years on May 21, 2010

Michelle & Geoff Thornell
Edmonton, Alberta, Canada
30 years on May 31, 2010

Laura & Lew Tillman
Northville, MI
35 years on July 26, 2010

Benefactors

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Dennis Rockford
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LOOKING AHEAD

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

Summer (current) 2010: Becoming A Parent With CF.

Autumn (November) 2010: Choosing The Right Caregiver. (Submissions due September 15, 2010) How do you decide what caregiver to use? Would you move to another location to be seen by a specific caregiver? Are there any tips you can share on how to avoid any pitfalls when choosing a caregiver?

Winter (February) 2011: Sleep Or The Lack Of It. (Submissions due December 15, 2010) Do you have trouble sleeping? Is getting a restful night of sleep difficult for you? Do you know any tricks for getting adequate sleep? Please tell our readers of your experiences.

Spring (May) 2011: Hobbies And Pleasure Activities. (Submissions due March 15, 2011)

ASK THE ATTORNEY

Information on Healthcare Reform

By Beth Sufian, Esq.

This column will discuss healthcare reform. Nothing in this column is meant to be legal advice about your specific situation or to suggest you change your insurance coverage. If you have additional questions, please contact the CF Legal Information Hotline at 1-800-622-0385. The Hotline provides free and confidential legal information to people with CF, their families, and their CF Center care teams. The Hotline is sponsored through a grant from the CF Foundation and can be reached by e-mail at: CFlegal@cff.org.

After over a year of debate in Congress and in the country about healthcare reform, the Patient Protection and Affordable Care Act became law on March 21, 2010. Most people still refer to the law as "Healthcare Reform". There are many parts of the new law that will help people with CF obtain health insurance coverage. Parts of the law start at different times. It is important to understand when each section begins. This column will discuss changes that will start in June or September of 2010. Future columns will detail other provisions of the law that take effect at later dates.

Some people have incorrect information concerning what the law will actually provide to citizens. The law does not create a national health insurance program. The law does not reduce the coverage that an insurance company can offer to policyholders. The law should make it easier for all adults and children with CF to obtain affordable insurance coverage in 2014. Insurance companies must sell an individual insurance policy to an adult regardless of health in 2014. Before then, it will still be extremely difficult for an adult (someone over the age of 18) to purchase an individual policy.

BETH SUFIAN

Below is a summary of the provisions of the law that are most likely to have the greatest impact on a person with CF and that go into effect in the next few months. Nothing in this article is meant to be a guarantee of specific coverage for anyone with CF. The summary is based on a reading of the law and limited guidance which has been issued by the Department of Health and Human Services (HHS), as of June 2010. The Department of Health and Human Services is drafting the guidance that will implement the law. The mechanics of how certain sections of the law will be implemented has not been decided. You can follow the issuance of guidance on the government website: www.healthcare.gov.

A. June 2010

1. Temporary National High Risk Pool

After June 2010, a Temporary National High Risk Pool (Risk Pool) will begin. The Risk Pool will provide coverage to adults who have been uninsured for at least six months as a result of a pre-existing condition. The Risk Pool will exist until 2014. At that time, insurance companies will no longer be able to deny coverage to adults with pre-existing conditions, and there will be state insurance exchanges that will offer insurance policies to those in need of coverage. The new Risk Pool should have premiums that follow standard insurance rates for the average person applying for health insurance. No premium price structure is available at this time. While efforts are being made to have the Risk Pool up and running in June 2010, there may be delays. Many people with CF will not be able to use the Risk Pool because, in order to be eligible, an individual must have been uninsured for six months and have no other coverage in place.

B. September 2010

1. Coverage for Dependents Under 26

"[This] law should make it easier for all adults and children with CF to obtain affordable insurance coverage in 2014."

Beginning in September 2010 (or upon renewal of the policy), children can stay on a parent or guardian's health insurance plan until the age of 26. If a child has reached a limiting age on a parent's policy but has not yet reached the age of 26, the child can return to the parent's policy in September 2010 when this section of the law becomes effective.

For example: Isabella turned 18 in April 2010. Isabella had reached the limiting age on her parent's policy and so lost coverage under her parent's plan. Isabella will be able to re-enroll in her parent's policy in September 2010. There is no requirement that the child under the age of 26 be enrolled as a full time student. Some individual insurance companies have decided to implement this section of the law earlier than September 2010. Other insurance companies will not implement this section of the law until the specific health insurance policy is renewed. Therefore, if the policy is not up for renewal until January 2011, the insurance company can wait to re-enroll children who have not reached the age of 26.

Young adult children under the age of 26 who are in need of insurance coverage should contact their insurance company to see when they can re-enroll.

Children under 26 who work for an employer who provides health insurance coverage will NOT be able to stay on their parent's or guardian's health insurance plan, once they reach the limiting age set out in the parent's or guardian's policy. However, in 2014 even if a child's employer offers coverage, the child can elect to stay on the parent's policy until he or she turns 26 years of age.

2. Individual Coverage for Children

Beginning in September 2010, pre-existing condition exclusions for children must be eliminated in all health insurance plans. This includes individual health insurance policies. This section of the law should mean that a parent who has a child with CF should be able to purchase an individual policy for their child. Details regarding this section of the law should be available soon.

3. Lifetime Caps

All new and existing health insurance plans must eliminate lifetime caps in September 2010. Some insurance companies say they will remove the lifetime cap only when the policy is renewed. Check with your insurance company to see when they will implement this section of the law. It will really help people with CF, especially those who have almost reached their lifetime cap and are in need of a lung transplant. A person must have a guarantee of coverage for a lung transplant in order to be placed on a lung transplant list. There are no exceptions. Most transplant centers need a guarantee for the cost of the transplant which is between $350,000 and $450,000. It has been important to know how close you are to your cap. Starting in September 2010 (or in some cases upon renewal of your policy,) there will no longer be a lifetime cap to worry about, and this should increase access to lung transplant for some children and adults with CF.

4. Annual Caps

Prior to 2014, annual caps on coverage will be restricted as allowed by the Department of Health and Human Services. It is likely an insurance company will have to follow rules set out by the government regarding their ability to impose annual caps on coverage. Starting in 2014 annual caps on coverage will not be allowed.

5. Rescission of Policy

Starting in September 2010, a health insurance company cannot cancel coverage due to higher use of benefits or a new diagnosis of a medical condition. The policy can be cancelled if the policyholder committed fraud in obtaining the policy or while enrolled.

6. Medicare Recipients

Adults with CF who are receiving Social Security Disability Insurance and Medicare will see new benefits thanks to the new law. In 2010, Medicare recipients will receive a $250 rebate if the individual reaches the donut hole in Medicare Part D. In 2011, the donut hole will be reduced by 50%. By 2020 the donut hole will be eliminated. For individuals with CF who are on Medicare, this section of the law will reduce the amount of money the adult has to pay for prescription medication. Medicare will also start covering preventative services and screening tests. ▲

Beth is 44 and has CF. She is a Director of USACFA. Her contact information is on page 2. You may send CF-related questions of a legal nature to: bsufian@usacfa.org.

SPIRIT MEDICINE

Why Bad Things Happen to Good People with Cystic Fibrosis

By Isabel Stenzel Byrnes

In the last few months, two friends of mine with CF have found themselves in terrible situations. One is getting listed for transplant after a long, hard fight. One declined precipitously and, before she could even be considered for a transplant, died. I don't understand what has happened. Maybe I'm an optimist, but in this day and age with CF, the "Pipeline", and all the new treatments, I somehow expect new medicines to control lung function decline; and if that's not the case, I expect people with end-stage CF to receive lung transplants. In this generation of so-called "controllable CF", it often seems we have so little control.

So, I find myself wondering why bad things happen to good people with CF.

Shortly after setting aside my first draft of this article, I received an e-mail from Kriss, a friend of mine who also has CF. She admires all the blogs, videos and Websites out there about people with CF who are able to run marathons, exercise, play in rock bands, and live productive lives. However, she also touched upon the theme of this article by saying, "I know people who have fought hard, been diligent, and *not* succeeded in surviving CF. I worry sometimes that our community and, especially, outside of the CF community, also need to know that you can do everything right, and *still* not thrive. I worry sometimes that we do not honor enough the memory of the people who have fought and lost. I have even seen in our community a judgment of those who are not thriving as people who were not strong enough, or positive enough, to overcome CF. In the spirit of awareness, wouldn't it be beneficial to show both sides of CF, and highlight the strength in both overcoming the odds and, sadly, the strength in not overcoming the odds?"

Upon further discussion, I decided to include Kriss' commentary in this Spirit Medicine. Kriss is a 33-year-old who does all her treatments, has good support, eats well but is still finding herself on disability, on the verge of being listed for transplant, trying to find quality of life amidst her chronic exhaustion and shortness of breath.

Kriss and I have seen how unfair CF can be. Some of us have very mild disease. Some of us receive lung transplants and live long, healthy, active lives. Some of us in the prime of life are suddenly struck down.

Last year, I was reconnected with a former camper named Emily. She was one of the few survivors of her peers, after a *Burkholderia cepacia* outbreak occurred at the CF camp 15 years ago. She was bright, beautiful and articulate, and found herself as a spokesperson and advocate for CF. As a robust athlete, Emily maintained her health by surfing regularly in Southern California. She became the spokesperson for the CF Surf Camps sponsored by Ambry. Then suddenly, early this year, her health took a turn for the worse and she ended up on a ventilator, fighting for her life for two months. At 27, she died.

> *"I can try to understand that in the suffering caused by illness, loss of function and death, we can seek a spiritual path to find meaning to this injustice."*

ISA STENZEL BYRNES

I'm still trying to make spiritual sense of all this. Emily and her family were devoted Christians. They went to church and prayed regularly. She did her treatments, ate well, exercised, and coped positively. I often ask God why Emily had to die so young, at the best time of her life. I have a feeling her family might have asked the same questions.

Emily, as well as so many of my friends, was such a good person. Why did she get the short end of the CF

stick? At my worst, I think maybe they did something wrong. But I knew them.

It is instinctual to judge – as a self-protective mechanism – and it happens too often in the CF community. I know firsthand how uncontrollable this relentless, cruel disease can be. My sister and I did chest percussion for four hours a day, each, until our hands bled, and we *still* couldn't stay out of the hospital or prevent the need for a transplant around age 30. On the contrary, I know some people with CF who don't do much for their health, and they somehow stay well.

Kriss rightly says, "Our challenge is not to judge how others handle their CF. Maybe the best service we can do anyone who has battled with significant disease and lost, is to appreciate their individual approach to their struggle and learn from it for ourselves. We need to remind ourselves that life is sometimes short and unfair, and to appreciate what we have, and respect what they didn't get enough of – time."

What else can we do? I've come to realize that I have no power over other people's paths with CF. It's a helpless but true fact. I can pray hard and long enough until I'm blue in the face but, really, people's lives will unfold the way they are meant to unfold. To some extent, the same can be true of my path.

All I can do, for myself, is find a way to make sense of it all. I can accept that sometimes things just unfold in ways that I do not want. I can try to understand that in the suffering caused by illness, loss of function and death, we can seek a spiritual path to find meaning to this injustice. A Buddhist teacher, Sogyal Rinpoche, says, "For us to survive on the spiritual path, there are many challenges to face, and there is much to learn. We have to discover how to deal with obstacles and difficulties; how to process doubts and see through wrong views; how to inspire ourselves when we least feel like it; how to understand ourselves and our moods; how to evoke compassion and enact it in life; and how to transform our suffering and emotions." It's a mighty task, but if we were chosen to have CF, perhaps we were chosen to explore these questions and ponder the answers, if there are any.

We can ponder what it means to 'get' CF. If we live our lives with CF as if it were some kind of punishment, what would that do to us? We would likely see the world, or God, or our fate in a negative light. Most important, we might think that there is something wrong with us—something inherently bad or evil about us. To me, the most disturbing biblical response is that which says there are no "good" people. According to gotquestions.org, "the Bible makes it abundantly clear that all humans are tainted by and infected with sin. We deserve to be thrown into hell at this very moment and every second we spend alive is only by the grace and mercy of God." This doesn't really work for me. It doesn't help me understand why my friends suffer.

> *"Maybe, we need to try not to focus on why, but on what we are going to do with our individual challenges."*

If I saw my CF as a curse, I would feel victimized, inflicted upon, maybe even oppressed. Those are heavy burdens to carry. My world would be pretty dark.

If I saw my CF as some kind of test, I would feel like a helpless recipient of struggle in the external universe with little control. I'd likely be exhausted. I'd feel anxious about the next hardship around the corner. But, on a positive note, Rabbi Shraga Simmons asks, "What does it mean to be tested? You have *potential*. The Hebrew word for test – 'Nisa', is the same as one of the Hebrew words for flag – 'Nes'. What's the connection? You hoist a flag; so, too, through being tested, we become hoisted to higher and higher levels."

If I saw CF just as something that is, then I give up trying to analyze it too much. I see CF as just happening, whether I want it or not. CF is just my life. It's easier to accept it, without judgment. Kriss sees CF as creating and forming who she is. She asks herself, "Could I be the person I am today without CF being woven into and through out my life? No. It is the good and the bad that has shaped me as a person, and without CF, who knows what or who I would be."

By saying 'bad things' happen to good people, I am implying that CF is bad. How one views having CF has great influence on how one lives their life. It's our own judgment that a short life is bad, yet I know many people with CF who pack 80 years into their 30 years of life. Kriss adds, "Maybe a 'good' life isn't just the time we have, but how we use that time."

If I try to see my CF as a mix of good and bad, then I feel better. Yes, CF has given me unimaginable possibilities, has exposed me to extraordinary people and circumstances, and given me unique maturity and perspective. Yes, I can say I'm living my life with intention, compulsive goal-setting, expression and love because

Continued on page 25

SPEEDING PAST 50...
Another Side To Parenting

By Kathy Russell

We all made it through another winter, but for many of us, it wasn't easy. Some of my friends didn't make it through and others struggled mightily…and managed to make it. I spent all spring battling pneumonia in both lungs. I ended up on oxygen continuously. Let me tell you that I don't like having a "leash", but I am so grateful that O_2 can make my life easier. I hope that O_2 - 24/7 will not be a part of my life forever, but if it is - such is life.

Being as sick as I was makes me realize, once again, how fortunate I am to have a wonderful husband who takes good care of me. Life without a partner would be so much more difficult. Thanks, Paul, for always being there for me. Now, on to the Focus topic of this issue: *Becoming A Parent With CF.*

Deciding whether to have children is a big decision and one that most of us do not make without thinking about it, long and hard. You have read my story many times so I won't reiterate it here. Instead, I will tell you of what some of my friends, who are parents of people with CF, have said about their children marrying.

This subject occurred to me when I read a post to an Internet support line that was written by the mother of an adult who has CF. In that post, she was extolling the virtues of her son-in-law and was expressing how happy she is that her daughter has such a wonderful husband. Another woman was writing of her son's marvelous wife. She is so happy that her son found a great partner and thinks that her daughter-in-law is terrific.

Both of those posts caught my eyes and reminded me of how my own mother loved my husband. We often joked that Paul (my husband) was my mother's favorite child. Mother was truly happy for me, that I had found a person who accepted me for me and didn't expect me to be a "super woman" or anything other than myself. She loved the fact that Paul and I loved each other and enjoyed spending our time together.

Some of my friends, who also have CF, and I have spoken of how fortunate we are to have good partners. The husband of one friend even had surgery on his sinuses to help stay healthier for his wife's sake! Each of us is sure that we wouldn't be around, if it were not for our partners and their care and concern for us. I know that definitely is true for me.

Now, don't get me wrong. We all appreciate our parents and what they did for us, but there comes a time when we want to make our own way in this world. It becomes time to leave the nest, break away and fly with a partner who is not one of our parents. I know that this is a difficult transition for some parents.

> *"We all appreciate our parents and what they did for us, but there comes a time when we want to make our own way in this world."*

I know of parents who just can't let go of their children. They feel that no one else can care for their son or daughter as they can. This may be true, but it doesn't mean that someone else cannot care for them just as well.

One mother whom I know says that her son-in-law is a "brick", a real solid man. She is so fond of him that she almost cannot express it. As her daughter has gone through various life-threatening episodes, her son-in-law has been there, solid as a rock, every step of the way, offering whatever assistance he can. His presence has been the difference in how quickly my friend (the daughter) recovers from each incident.

Another mother, whose daughter has died, was so happy that her daughter was able to marry before she died. Even though the marriage was short, it was a true love-match, and for that she was very grateful. We have talked of this, at some length and she is so happy that her daughter could know

KATHY RUSSELL

that kind of love and could share her love with the right person.

To illustrate what I am talking about, I am inserting two stories that were written by mothers of people who have CF about their children's partners. The first is written by Deb Pence:

"For richer, for poorer, in sickness and in health, to love and to cherish; from this day forward until death do us part." Before my children were diagnosed with cystic fibrosis (CF), I would hear those words at a wedding and I would look at the bride and the groom and would think to myself that they have so much time ahead of them before those vows would ever become meaningful in their lives.

I found myself becoming a single mom at the age of 32, raising my son, Kevin, just turning 5 and my daughter, Olivia, who had just turned 2. I was young, full of energy and had big plans of making their lives so full and meaningful. CF had not yet become a part of those plans, at least not on my list.

On June 1, 2000, when they were 12 and 15, CF penciled itself in and moved into my home uninvited. Things really changed, including the meaning of that vow. Of course I wouldn't think of it again for at least ten years.

I've learned so much about CF in that time and it's a continual learning process. I experienced feelings and emotions I never thought possible, and in those 10 years, while we moved through puberty and the "teen" stage into young adults, there were times I thought I might lose my mind. I finally realized that teenagers are why animals eat their young. (Humor helps when dealing with the teen years)

My mind, however, remained intact through it all, but I always worried about who they would wind up with in life and even if they would find someone to accept them, CF and all. As with most parents, we always want the best for our children and finding someone in our children's lives, as mates and partners, seemed so much easier for my friends' children. CF is a big deal breaker, when it comes to love.

After 20 years of raising them by myself, I knew how much I loved them, and as a parent all you want is for them to find someone who loves them even half as much as you do. You know their hearts, you know their needs, and you want them to find someone to accept every part of their beings.

My daughter would always ask me who would want someone like her who is broken and flawed. I would cry and tell her those flaws are what make her unique and beautiful and that the man who gets her will be lucky to have such a kind, loving person with an enormous heart. Over the years no one truly appreciated her or understood the trials - no one until now! Thank you, Kurt, for loving her the way she deserves to be loved.

> *"I always worried about who they would wind up with in life and even if they would find someone to accept them, CF and all."*

My son had a couple of longer term relationships but, somehow, as a mom I didn't feel those were the ones. Those few had not loved him back the way he loved - with all his heart. The ending of those were difficult for him, and I prayed he would find that one who appreciated his sensitive side and could see how he understood the real value of life.

My son had been dating my daughter-in-law, Samantha, for a brief time before I met her. When I did meet her, I realized exactly what he saw in her and it felt like the weight of the world was lifted from my shoulders. I knew that he had found the one who loved him as much as, if not more than, I did.

She is strong and beautiful and although she tells him he is overly sensitive and emotional, I know that when he sends those flowers to her at work on those special days, she is glad his heart ticks the way it does. During those dating years she would always stare at him and tell me how beautiful my son was. Today, as his wife, when I ask her if she still thinks he is beautiful, she looks at him with those eyes and says, "Yes." Thank you, Sam, for loving him as I have.

I believe that when a person with CF finds the right person, it's a love that goes beyond, "I take thee to be my lawfully wedded husband or wife", it exceeds "do you promise to love, honor, cherish and protect", and it's truly about, "in sickness and in health, to love and to cherish; from this day forward, until death do us part." I saw that in my son and daughter-in-law on their wedding day, and it was that day that I realized the true meaning of that vow.

For those of you who haven't reached this stage of life with your child with CF, I pray that God will be as kind to your hearts as He has been to mine, that your lives will be blessed with people in your child's life who love them as much as you do and that you will raise them with the thought that "God won't make a mountain they can't climb."

The second story was written by Teresa Hook-Klaus:

My daughter Shannon is 30 years old. She was diagnosed with cystic fibrosis (CF) when she was five months old. She married her junior high sweetheart two months after graduating from high school. They divorced after seven years of marriage.

Shortly after that, she reconnected

Continued on page 16

FOCUS TOPIC

BECOMING A PARENT WITH CF

Sweet Music

By Jeanie Hanley, MD

JEANIE HANLEY

Respiratory treatments are a way of life for us. For most, there's no getting around them if we want to maximize our health. I believe that incorporating them into the lives of our children is critical for their adjustment and to ours and, rather surprisingly, this can also bring a wondrous symphony of benefits.

I have two teenage girls (15 and 17) and an adult son who just turned 21 in August. My husband and I have been married for 24 years and, luckily, have been on the same page as we navigated the different ages of our children and learned how to parent as we went along. Something I was always concerned about was the effect of my CF on the social lives of my children. I've always performed the Vest and my nebulizers in the dining or living room of our home. This approach was just more convenient and comfortable and I was able to keep tabs on the beat of the household. Also, I had decided early on that I wasn't going to hide myself in a room doing treatments. As a consequence, a wealth of benefits, mostly unexpected, have followed.

When my kids were younger, simply my availability to hear what happened after school became a ritual. Sitting there with a nebulizer stuck in your mouth, you become a good listener! I tried to complete my afternoon treatment soon after they came home or whenever I came home - sometime before dinner - whenever possible. Once I began part-time work, this became easier and as they became teenagers and their workloads skyrocketed, this time after school became essential to help them with homework. At times, one child would finish while the others would watch a game or take part in some other noisy pastime. The Vest and compressor/nebulizations effectively created a wall of white noise that drowned out these extraneous noises, thus allowing for better concentration. I obviously cannot be sure about this, but our dining room may be the only "formal dining room" made very informal by textbooks, computers, papers, pens, etc. littered across it that are absent by day, and reappearing early every evening.

The other unexpected benefit is the reaction or lack thereof, from my kids and their amigos when they come over. I've always encouraged the kids to bring friends over or have parties here so that I could meet and get to know them. This is key to understanding much of their teenage interactions and behaviors with others. The surprising outcome of this is that my kids are unfazed when my active treatments are underway, so their friends are similarly unfazed. Either they were just being really cool or, after they realized that I could take a break and talk or that it was simply, my family's "normal", which took all of a few seconds, then they went about playing pool, video games, piano, singing (lots of drama kids around), or other forms of entertainment. Only a couple of times were two kids a little shocked at seeing me with what appeared to be tubes running into my chest, vibrating the whole upper body and a plume of smoke emanating from a strange device in my mouth. Come to think of it – it's amazing that there are only two instances!

Another advantage of being open about CF and the required treatments is awareness. There is now greater awareness of what CF is among my kids and their friends, many of whom have tried the Vest and asked insightful questions. They also have been

inspired to write about it, give slide presentations during biology classes and, in one instance, taught the science teacher about CF genetics. Just this summer, my son, who has finished his third year of college, was accepted into a research internship with a chemistry professor who studies dysfunctional protein processing such as occurs in CF cells. Very importantly, they have participated and/or organized fundraisers. Over the years due to my children's leadership in Great Strides Walks, and of local schools in the CFF Cure Finders Program, they have raised over $15,000.

> "My kids are unfazed when my active treatments are underway, so their friends are similarly unfazed."

As my darling teenagers mature, I still try to be sensitive to possible changes in viewpoints and any embarrassment that my illness may cause. I believe that we communicate well and, if any new kids are coming over, and I must do my treatments during their get-togethers, I will check with them just in case. They have always responded with some form of: "If they can't handle it, then tough." or "They don't care." or "My friends are fine with whatever." which always is sweet music to my ears. ▲

Jeanie is 48 and has CF. She is a physician who lives in Los Angeles with her husband, John, and three kids, Kevin, Maria and Jessica. She is a Director of USACFA. Her contact information is on page 2.

CLUB CF ONLINE

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

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

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

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

Announcements

VOLUNTEERS NEEDED FOR STUDIES AT NIH

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

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

We are looking for individuals with cystic fibrosis who previously participated in NIH studies. If you have taken part in an NIH study, please call the toll free number: 1-877-644-5864 and select #3 on the menu; or send an E-mail to: barnesp@nih.gov.

Becoming A Parent With Cystic Fibrosis Is Not Easy

By Steve Gonzalez

Hello. My name is Steve Gonzalez. I am 45 years young and I have CF. I was diagnosed at eighteen months of age. I have been happily married to my wonderful wife, Bridget, for the past 16 years. We have a beautiful son, Douglas. He turned 9 on June 24. We live in a nice home, in a great neighborhood (Rockville Centre) on Long Island, New York.

I tell you all this because the thought of reaching adulthood and being able to have a family when you have CF once was thought of as unreachable. Thanks to all the great new medicines and advancement in treatments it is not only possible to have children, but it is possible to have the full life we all dream of having. Becoming a parent with CF was not an easy process by any means.

Bridget and I started our journey to begin our family in 1997. As a male with CF I was sterile so we could not go the conventional route. We opted for In Vitro Fertilization (IVF). This was an extremely difficult and frustrating process, not to mention very expensive. On my end I had to administer daily hormone shots to Bridget. On Bridget's end she had to receive them. This was very difficult for her as it is a very involved and lengthy process, which sometimes caused her much pain and discomfort. This also involved many trips to the In Vitro clinic and lots of blood work, ultra sounds and sonograms. Our first attempt of going through this was unsuccessful. We thought long and hard and decided that, despite the emotional roller coaster we went through, we were determined to have a child and start the family we both wanted. Well, after another cycle of In Vitro we were still unsuccessful. That was quite a sad time for Bridget and me. What to do next?

You know the expression "If I knew then what I know now"? Well,

STEVE GONZALEZ

now I know - adoption! We decided to pursue this avenue so we could fulfill our dream of starting a family. I got the name of an adoption attorney from my CF Center. Some other patients worked with this attorney and had much success. What a home run this was! We started working together in March of 2000, and in June of 2001 our beautiful son, Douglas, was born and our dream of becoming a family was now a reality!

Now the fun begins. Raising a child (or children) is a time-consuming task that requires your utmost devotion and love, and is a sometimes overwhelming task. For me, it meant that I really needed to take care of myself. My plan is to be around for a very long time so that I can watch my son grow up and enjoy his life with him.

When you have a child your life changes, and everything is now centered around your child. If you are a parent with CF, then you have an even greater challenge. You need to be very disciplined so that you can devote the time it takes to care for yourself, and also devote the time it takes to care for your child and family. As a CF patient the older you get the more things you encounter. You need to keep yourself as healthy as possible so that you can overcome any unwanted situations that arise, (IV treatments/hospitalizations/diabetes, etc...) This is not easy as, the older we get, the more we need to take care of ourselves.

This is especially hard for me because I am a very active person, and I am very involved in my son's life; school activities, soccer, football, basketball, baseball - you get the picture. I am a very involved parent and I wouldn't have it any other way. When you have children, you need to be able to devote much of your time to them. So how do you devote the time to keep yourself healthy and well?

Fortunately for me I have the great motivator - my son, Douglas. I remember the day he was born; as I was holding his little body in my arms, I made a promise to him, to Bridget and to myself that I would do whatever I needed to do to always be there for him. It was such an overwhelming feeling of joy to hold him; I knew that nothing would stand in my way, and it hasn't. I really stay on top of my health. Have there been some

> We thought long and hard and decided that, despite the emotional roller coaster we went through, we were determined to have a child and start the family we both wanted.

rough times over the past nine years? Of course there have. I look at it like this: if I have to have occasional IV therapies to make me feel better and improve my quality of life, then so be it. It is not the end of the world to me. If it is going to prevent infection and keep me going, then all the better.

Being a parent has been the single greatest accomplishment of my life. I would not trade it for anything. Sure there are many trying times and frustrating moments, but the joy of having a family is one of the greatest feelings you can have.

So what does the future hold for my family and me? Well, after we successfully adopted Douglas, we decided to wait a bit before we began the journey for a second child. You know how that goes. One year turns into two, then three, and so on. Finally, when Douglas was almost eight, we started the process again. This time it has not been as easy as the first time, and we are currently still in the midst of pursuing adoption of our second child. The decision to adopt a second child was not an easy decision to make, but we considered all the factors that were pertinent to us - age, time, money, health, and a sibling for Douglas. Bridget and I realized that the thing we wanted most was to increase our family by one more. Although it seems like we waited a long time before making this decision, we realized one thing - if there is something that you really want in life it never is too late! ▲

Steve hopes that sharing his experiences and journey has been helpful. If anyone wishes to contact him, his phone number is (516) 992 - 2481, and his email address is: drummersjg@hotmail.com.

In These Trying Times

We know that some people may be going through a hard time with their employment and/or their medical insurance. We are letting you know about a program Genentech is offering called Pulmozyme Access Solutions. Remember, even when times are hard, it's important to keep taking your medication as prescribed by your physician. Focusing on your health is the best way to be there for your family.

Pulmozyme Access Solutions is Genentech's commitment to cystic fibrosis patients. They are here to help find a way for you to get the Pulmozyme your doctor has prescribed.

- **Do you have questions about your insurance coverage for Pulmozyme?** They can help you navigate benefits, coverage or reimbursement issues.
- **Have you recently started Pulmozyme or changed insurance companies?** The StarteRx Kit is a free, 30-day supply of Pulmozyme, nebulizer and educational materials provided to patients initiating therapy while insurance coverage is ascertained.
- **Do you need help with your co-pay for Pulmozyme?** They can refer you to independent, non-profit organizations that provide co-pay assistance and help you with the application process.*
- **Are you uninsured, has your insurance company denied coverage for Pulmozyme or have you met your annual or lifetime insurance cap?** Genentech Access To Care Foundation provides Pulmozyme free of charge for eligible patients without insurance coverage.

If you answered "yes" to any of these questions, Genentech specialists can help you or someone you know. Call **(800) 690-3023** from 6 a.m. to 5 p.m. PT, Monday-Friday, or visit **PulmozymeAccessSolutions.com** anytime. Check them out if you are in need. They are here to help us out.

*Genentech cannot guarantee co-pay assistance once you have been referred by Pulmozyme Access Solutions. The independent, nonprofit organizations to which patients are referred each have their own criteria regarding eligibility, including financial eligibility. Genentech does not influence or control the operations of these independent, non-profit organizations, but Pulmozyme Access Solutions can help you navigate the process of seeking co-pay assistance by referring you to an appropriate organization and by assisting with the application process.

The Joy Of Being Called A Mommy

By Bracha Witonsky

I sit here excitedly about to write an article for CF Roundtable about being a parent and having CF, and on my right sits my 9-year-old daughter, Shaina, singing at the top of her lungs some of the songs she learned last year in school. She has a cute, bubbly smile, her hair is in a bun and she is dressed in aqua from head to sandals.

There is so much to tell you. I literally could write a whole newsletter on this topic alone. I will do my best to express my joy that I have been able to be a mother of two beautiful gems. Shulamis Zahava is sitting in the living room, also dressed in aqua – with her long straight hair pulled back with a headband. She is 10-years old. My girls are 14 months apart in age.

I got married at 21 to a wonderful "mensch", Yonason. Before we celebrated our first anniversary, our first gem was born, Shulamis Zahava, who made me into a mother — a dream that I have always had. The pregnancy was complicated, but the real challenge was the time after pregnancy — raising a child. I constantly need to be ready at any time, day or night, tired or not tired, feeling good or feeling lousy, for my child. My child, like any other child, needed a mommy to care for her at all times.

Since I kept my CF a secret throughout my life, I had to make a decision about what my husband and I would do with our family – one child at the time. Would we continue to keep it a secret, would we tell others, and would I allow myself to do my treatments in front of my baby — or only do them when she's sleeping? We had to figure out the right balance in whom and how much we would and would not keep secret. The secrecy factor would determine how traumatic our everyday life would be.

We felt, for the benefit of ourselves and our child and future children, it would be best to live life as "normal" as can be. So, to our baby at the time, things would appear to her as if we were a regular family. All the treatments that I did, like swallowing pills before I ate, checking blood sugars constantly, using my insulin pump to administer insulin, doing my nebs, vest or hand therapy (a Physical Therapist used to come daily) and all the rest of the stuff I do continuously to keep healthy, I would do, just as normal as a person without medical needs. I was worried that my baby seeing me getting treatment, for example, might view me as if I was getting hit and I was in pain and uncomfortable having to force myself to cough. But, honestly, the fact that we agreed to not keep anything a secret from our child since she was born, made all my treatments appear to her as if all mommies need to do that, and it's actually normal.

BRACHA WITONSKY

Sometimes she would play house with her dolls and would do CPT treatment on her doll and ask her doll to give a big cough, and give the doll a tissue to spit into!!! Then with the blessing of G_d, 14 months later, our second gem was born. Yes, fitting in my treatments and caring for two small preemie babies was a huge challenge. I thought the hardest part would be the actual pregnancy, but being a very devoted mother to two, 24/7, was the bigger challenge.

We do things a little differently from other families, to make sure everyone's needs are taken care of.

We train our kids at an early age to help do chores around the house. Never underestimate what young children can do. Kids like to get involved when you allow them to. It empowers them, and makes them feel great. (My kids cook supper frequently and can make soup and chollent for Shabbos.)

As my girls got older, like the age where they have play dates, my children would come home with the smartest questions, "Mommy how come that girl's mommy does not test her sugars before she eats, like you do?" or "Mommy this one's mother didn't cough once the whole time while I was there." I gave them simple true answers that were on their level. I know for a fact that there are for sure mommies that have CF, other mommies that have just asthma, other mommies that have just diabetes — the list can go on and on. With all this in my mind, I told my children that G_d made everyone, and no two people are the same. We all need to go to our own doctor and get examined. According to what the doctor sees, he tells each person what they should do to keep healthy. He also tells us how often we need to be seen. So, for me, I need to do certain treatments and swallow pills daily. For you, your doctor says you need to take one vitamin daily.

Generally speaking, I did not talk about my CF to other people other than the obvious few family members who already knew. I acted very openly in my house with my family, when doing any treatments or taking my medicine. My children picked up on not yapping to other people about what I need to do to keep healthy. They realized that regarding this topic, we do not talk about it to people other than our family. One thing that we felt was very important was not to tell my children, "You can't tell anyone what Mommy has" or "Don't tell your friends that Mommy takes medicine"! My kids, on their own, see that my CF is something private. We don't openly blabber about it.

However, I do speak to my girls' principal and teachers every year, to let them know about my CF, only out of necessity. I am careful to tell them this information after only a few weeks of school, so that the teachers can get to see my kids as any other kids and get their first impressions of them, before they know who they really are. I want them to know that there is a possibility of their mom going into the hospital for long periods and what their mom having CF entails for my kids. This way, they understand my girls throughout the year. There may be times when, for instance, my kids won't have their snack bags or their hair may not be neatly brushed, because I am hospitalized. They should know in the backs of their minds – maybe something is going on at home and some extra TLC would be very much appreciated.

My children are extremely mature for their ages. When I come home from the hospital or if I am home and really not feeling well and I ask them to get me something, they will run as fast as they can and not ask me any questions. The minute I ask them to do something for me and they act like any other kids their ages - whine and complain - then I tell myself, "Bracha you must be feeling better!"

When I am feeling well, I give to my children 200% of what a "regular" mother without medical needs does for her children. I know that and so do my kids. The reason I do that probably is to tell my brain not to feel guilty for the times I can't mother my children like other mothers do.

Now that my children are getting older, they are starting to realize that we are not alone! Other families have parents or children with some sort of medical needs and also are restricted from certain foods or activities and also require medicine. This helps our children understand that G_d created different people with different needs. We need to fully accept our own unique situation, whether we are happy with it or not. This is the situation that G_d gave us, and we will take it and live life to its fullest, to the best of our abilities.

In my religion, being orthodox Jewish, we like having large families. Being able to be called a WIFE and a MOMMY, and being able to say, "I am a full-time mother to my CHILDREN," is the biggest blessing to me. Yes, I would love to have more children and, especially, a boy. But my health is the most important thing to me and my family. I do realize that putting my body through more pregnancies would put me at risk. So, I thought of maybe adopting a baby boy. But having said that, the challenge is not really so much the nine months of being pregnant, it's all the years of raising them and caring for them all the time. So, adopting at this point will not be my answer, being that I thank G_d I already have two precious kids who take every ounce of my energy!!

Here are some day-to-day tips on how I manage taking good care of myself and fitting in all my medical treatments while juggling my family and being there for them.

1. I try to wake up ten minutes before the girls wake up to have my cup of coffee and have a few minutes of peace before the yelling and screaming starts – "Where's my sock?", "I can't find my shoe!", etc.

2. After I come back from the bus stop, around 8:30 AM, I take care of myself for about an hour. Eating breakfast, drinking shakes, doing treatments, the works! Now, with the summer weather, I sit outside on a lounge chair and sit in the sun. For

Continued on page 25

over the Internet with Jason, whom she had known in high school and who was then in the Air Force stationed in Korea. They communicated online and through web cams for almost a year, getting reacquainted. Jason also was divorced and had a daughter and a son. As the months went by, they grew closer and closer, and it seemed as if the friendship was growing into something much deeper.

When he returned from Korea in 2007, Jason was to be stationed at Whiteman Air Fore Base in Missouri, a little over an hour away from where Shannon was living. They agreed to take things slow at first because of his children and Shannon's health, but it sure was love! Shannon and the children love each other. They've all handled it when Shannon has been in the hospital or when Jason went overseas for three months or out of state for training. They are a family.

On May 29, 2010, Shannon and Jason were married at a winery not far from their home. It was a beautiful outdoor ceremony. Shannon walked down the aisle on her father's arm to Etta James singing "AT LAST", with Jason looking at her with such love in his eyes that the tears of joy started for many in the audience - especially me. When they said their vows that they each had written themselves - talking of the love, friendship and respect they have for each other – the tears were pouring down my cheeks. Shannon also spoke of the joy that the children bring her, and her commitment to being the best "bonus" mom possible to Jason's children.

As we got to the end of the aisle after the ceremony, I was sobbing. It was all just so "right". The first thing I saw was Payton, Jason's 14 year old daughter, in Shannon's arms also sobbing. Then Jason and 10 year old Gavin joined them for the first official group hug with all four of them. I, of course, kept right on sobbing, and had to walk on a little way alone to compose myself.

I've never been one who is publicly emotional. I think most moms of children with CF learn quickly to hide the tears and fears from others. The bathtub or shower with the water running high is my favorite place to bawl like a baby. Since Shannon's birth and diagnosis of CF, I've always been supportive of Shannon's desire to live her life, including decisions I disagreed with. I am sure her father would say the same thing. As with most teenagers and young adults, Shannon made her fair share of bad decisions – some to do with dealing with CF – and some with normal life. (Her brother and sister made the same mistakes and they don't have CF.)

Most all children grow up and leave home – children with CF included. I've been mother of the bride three other times, including Shannon's first marriage. No empty nest syndrome for me. I was happy for them to start their lives as adults, and I was happy for me! Unlike any of my children's previous weddings, that were all very large beautiful church weddings, I honestly can say that I've never felt such peace settle over me as I did at the winery. The emotion, pure joy and beauty that I witnessed between Shannon and Jason was a spiritual experience.

Nobody with CF has an easy life. At some point every person with CF starts getting sick and, gradually, sicker. There are peaks and valleys. It varies with each person – but the one sure thing is, it will happen. We parents learn to be strong in front of our kids – and sometimes with each other. I think we all fear that if we lose control emotionally, we'll never get it back together. Watching us be strong, caring for and advocating for our children, teaches them to be strong.

I love, admire and respect Shannon. She is an amazing young woman. She's not always had it easy. She doesn't remember trick or treating as a kid – she always was sick on Halloween. She was homebound her senior year of high school due to being so sick. She had a marriage (and a husband) that failed. She's been in and out of the hospital and on IVs so often in the last few years that I've lost track of the number of times. Yet she keeps right on living and loving.

My new son-in-law, Jason, is just as amazing as his wife. It takes guts to fall in love with a person with CF. He's a medic in the Air Force. He chose to embrace Shannon, rather than run away from dealing with her health. He is an amazing father. I have so much respect and love for him – he loves my daughter unconditionally, knowing full well that it won't always be an easy road, but they will make it through whatever comes at them together.

What more could a parent want for a child?

Stories like Teresa's and Deb's aren't always the case. I have known parents who were unable or unwilling to let their adult children who had CF go off on their own. They interfered with the relationships that their children had and even with their subsequent marriages. I remember talking with one woman, whose husband had CF, and her mother-in-law was making life miserable. The mom was constantly checking with her son to be sure that he had taken a medicine or done a treatment, and if he had missed one she blamed her daughter-in-law. That couple eventually divorced. It wasn't that they didn't love each other or that the wife couldn't take it; it was just a case of too much mother in their life. The mother got her son back, but he died subsequently and I believe it may have been from a "broken heart".

Happily, I can say that is a rare case. Most parents adjust to the idea that their children, even those who have CF, will grow up and go out on their own. They may miss some meds and/or treatments, but they will survive and they will learn what they can and cannot get away with. Parents just have to learn to bite their tongues and keep out of their kids' lives. If the parents have done a good job of raising their kids to "live" with CF, rather than raising them to be invalids, those kids will do well. Be happy for them.▲

Kathy is 66 and has CF. She is a Director of USACFA. Her contact information is on page 2.

PHOTO BY MICHELLE COMPTON

Womb

You liar
You cheat
You thief
You warped mass of genetic inaccuracy
Killing all without regard

You have morphed into every aspect of my life
Scarring
Violating
Poisoning and infecting
A painful surprise around every corner

You took from me my options for
A normal life, a natural family;
freedom from anguish
You took my hopes for a child -
My baby

My baby.

– Melissa Thompson, 2005

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

To learn more about us and view more images in the collection, please visit our website at: http://www.thebreathingroom.org

FROM OUR FAMILY PHOTO ALBUM...

DEBBIE AJINI AND CATHY VANACKER AT A TRANSPLANT FUNDRAISER CAR SHOW FOR DEBBIE IN CLINTON TOWNSHIP, MI.

CLOCKWISE FROM LEFT: SHULAMIS ZAHAVA, BRACHA, YONASON AND SHAINA WITONSKY.

MICHELLE AND GEOFF THORNELL ENJOYING THEIR 30TH WEDDING ANNIVERSARY TRIP IN COSTA RICA IN STYLE!

JOHN, KEVIN, JESSICA, MARIA AND JEANIE HANLEY AT CHRISTMAS 2009.

BRIDGET, DOUGLAS AND STEVE GONZALEZ ON VACATION IN MEXICO.

FROM LEFT, ANNE WILLIMAN, DAUGHTER, AMY, 30; SON JOE, 26; HUSBAND, JON; AND SON JESSE, 18.

NOEL AND JANICE WHITE IN THE NC MOUNTAINS.

I'm a Hero of Hope, Who is Your Hero?

By Janice White

My name is Janice White, or Jan to those that know me best; and I am a 48-year-old woman living with the chronic illness, cystic fibrosis (CF). I am a *Hero of Hope* Living with CF, and I want to share my story with all of you, as we are all heroes

In terms of my career path, I have a Masters of Education in Speech Language Pathology and have worked for many years in this field, including with people who suffer from brain injuries and with the elderly living in long-term care facilities. Due to infection control issues, I no longer work with patients and now work for the state of North Carolina, where I help ensure that governmental policies are going to best benefit these patient populations.

> "I work out at the gym regularly, but I also like to take walks with my dogs to keep exercise fun."

JANICE AND NOEL WHITE WITH THEIR DOGS, COCOA AND PHEOBE.

As for my experience with CF, my story is actually very different from most as I was not diagnosed until I was nine years old. My older sister was misdiagnosed for three years, and her eventual diagnosis became the catalyst for my being tested. Sadly, she passed away at the age of 17. None of my three older siblings have CF, though one of my brothers is a carrier.

In 2003, a new chapter opened up for me and I received a life-changing bilateral lung transplant. In January, I celebrated my seventh anniversary with my new lungs! I know many of you can relate to the challenges I faced.

Before my transplant, a typical day included nebulizer treatments and chest percussion therapies multiple times a day, many medications and an exercise regimen. Since the transplant, I still focus on being compliant to my health routine, but the improvement in my health has allowed me to make some adjustments. I now take daily transplant medications and maintain my exercise program, which is a big part of my life. I work out at the gym regularly, but I also like to take walks with my dogs to keep exercise fun. Listening to my body and my doctor is imperative, and I take the time for rest and relaxation when needed.

I also work with the Carolina Donor Services, a procurement agency on the east coast, and the Sweet Melissa Fund, an organization at the University of North Carolina, which raises money for transplant patients. I enjoy helping with fundraising for the Brain Injury Association of North Carolina, and lately have become active with the Amyotrophic Lateral Sclerosis (ALS) Association, an organization dear to my heart as a close friend recently was diagnosed with ALS.

As a post-transplant patient, I feel it is very important for me to give back to the CF community and to other people in need. Now that I have the energy, I want to use it to help those who don't have the same luxury.

When my sister passed away, I was in the hospital chapel where a placard on the wall read, "Never put a question mark where God has put a period." Those words resonated with me, and ever since then I try not to question what I have been given. It's not always easy, but it is something to work toward.

Thank you for letting me share my story with you and I encourage you to nominate the Hero of Hope in your life!

For more information go to: http://www.heroesofhope.com/heroesofhope/docs/hoh_nomination_form.pdf to access the nomination form.

the way. We went sea kayaking and explored the coast line. We also met people from all over the world. It was a fascinating country to visit and, hopefully, we can go back some time.

We also made a couple more visits to Mexico, went to Tucson, Boston, San Francisco, St. John's, New Foundland, Victoria and Vancouver and continued camping and skiing in Alberta and British Columbia. On all these trips my health was never more than an occasional inconvenience. I would always try to do a "tune-up" prior to a big trip, scheduling a couple of weeks of IV therapy beforehand. Whatever I was doing seemed to work. Luckily, even though CF has robbed a lot of my health, I am still considered to have "mild to moderate CF". I am able to do the minimum treatments while I am on vacation. I take all my medications, have my husband for extra physio if I need it, but I am able to leave my compressor at home and take a vacation from nebulizers for a couple of weeks. I wouldn't necessarily recommend this for everyone, but it has not seemed to harm me.

This year we planned another trip to Costa Rica to celebrate our 30th anniversary. Unfortunately, in February, I developed a deep vein thrombosis in my groin and leg secondary to my IV port. We were very concerned about resolving this before our early May trip. I was started on heparin shots and eventually the port was removed. Since I no longer had IV access, doing a pre-trip IV antibiotic tune-up was just not convenient. Since I wasn't actively ill, my doctor and I agreed that I would do two weeks of TOBI® and Cipro before I went. Two weeks before the scheduled trip, I came down with a good old fashioned cold. My chest was very congested and I began to fear that I would ruin the trip. I came around and by travel day, while I was still coughing a lot, I felt much better.

Off we flew and embarked on a wonderful vacation. We met my husband's sister and her husband, who had gone a week before us, and spent a few days with them in Montezuma, my favorite place in the world. We went snorkeling, hiking, bird watching, visited butterfly gardens and generally had a great time. We left them and struck out on our own to explore other parts of the country. More jungle hikes, kayaking through mangroves, visits to nature reserves and long lazy days on the beach were at hand. The temperature was very hot– always over 30 degrees Celsius - and very, very humid. I was having a lot of difficulty breathing with any exertion. My Ventolin inhaler was never far from hand. Still, I was having a blast.

We traveled inland to a volcano resort that is rated the number two luxurious resort in the country. It was time to pamper ourselves for our anniversary. This resort had everything the nature nut would want. There was an aviary, butterfly gardens, hummingbird garden, snake and reptile house, frog house, jungle cats, monkeys, waterfall hike and more. The temperature here was actually cool at 16 degrees C, but still very humid.

Our room was something to behold. It had a king-sized four-poster bed, fireplace, sitting area, balcony with a hammock, rocking chairs and a hot tub. The bathroom was bigger than my living room at home. It featured double sinks, a waterfall shower, a regular shower, a waterfall and another hot tub! Live plants and trees were everywhere inside the bathroom! It was something out of Lifestyles of the Rich and Famous and we were lucky enough to get a very good deal as we went in the off-season. It was truly magical and a great anniversary present to each other.

The morning we packed up to leave the resort to go back to the beach for our last few days, I woke up very early and, while out on the balcony, coughed up a couple of tablespoons of bright red blood. While this is not really unusual for me, I was worried about it, seeing as how I was in a foreign country and my travel insurance would not cover a hospital stay. As the day wore on, I continued to cough up blood to the point where we were seriously considering contacting the airline. I kept my husband and myself calm and we decided to give it overnight to see what happened. I had discussed medications with my doctor prior to the trip and he urged me to take along prednisone and antibiotics. I figured that there must be a lot of inflammation in my lungs considering the amount of Ventolin I was having to take and so started on 50 mg of Prednisone that evening. I also began 750 mg of Cipro and discontinued my blood thinners. I kept an icepack on my chest and the bleeding seemed to stop. We were able to go out for dinner that evening without problems.

The next day, I had only had a few small episodes of blood, so I continued with my treatment. We lounged around by the pool with me trying to avoid the sun, because of the Cipro. I didn't feel sick at all and the bleeding stopped, so we decided we could wait the two days and go home as scheduled. Even though it all turned out okay, it was a scary incident. I am so very thankful that I came prepared and knew what to do and that I had discussed treatment options with my doctor before the trip.

I just got home last night and believe it or not, am flying to Phoenix on Friday! It will be a quiet, long weekend for me as I don't want to risk golfing in the heat with the rest of the crew. I will sit by the pool with a good book. Oh, and I will also be going to Niagara Falls in a couple of weeks. I'll bring my Prednisone! ▲

Michelle is 51 and has CF. She and her husband, Geoff, live in Edmonton, Alberta, Canada.

Parenting Isn't for Sissies

By Anne Williman

> "Kids not only have tons of energy; they suck every last ounce of yours. That means you have to have a plan to preserve yours."

When Jon and I got engaged, one of the things we discussed in detail was the issue of children. Oh, we both knew we wanted them but there was one big problem: my CF.

Back then, a pregnancy was pretty much considered suicidal for me. So the obvious answer was to adopt. We just didn't have a clue how difficult that was going to be, especially since we wanted a healthy baby.

The private adoption agency that we went to had a policy that you had to be married three years before applying. So as soon as we hit our third anniversary, I turned in the application. It would be another three years before we received a perfect five-month-old — our daughter Amy.

She was so perfect that we wanted more. This time, we went through an attorney who did private adoptions. It meant more waiting, but three and a half years later, we were placed with a two-month-old baby whom we named Joseph.

Having our two young children was challenging but, oh, so rewarding. And it wasn't long before we knew we wanted a third baby. This time, it was even more difficult, and we pursued a number of different options, including foreign adoption, special needs, and private adoption.

At one time, we even seriously considered a baby with CF. I wondered if she would be too much for me to handle; my own health care was plenty to manage, along with our two kids. Yet we felt we would certainly understand her unique needs and we felt willing to proceed. I even had a name picked out: Christy Kay. But it wasn't meant to be—my CF doctor was concerned about cross-infection and would not approve the adoption.

More years passed, and time after time, we would get close, only to have an adoption fall through, sometimes at the last minute. In all, I think we had about 10 babies that we somehow didn't get. It was heartbreaking, but we kept going.

Finally when Amy was 12 and Joe 8, our third and final baby arrived through another private adoption. We named him Jesse, and he was only five days old when we got him. Our family was complete.

Now, all of our children are adults (Jesse just graduated from high school.) Raising them was, at various times, the best or the worst part of our lives. If I were considering parenting today, I would give a lot of thought to several things.

Am I willing to be seriously unselfish, putting my child's needs before my own, on an ongoing basis? Here I'm talking money (For years, I literally never bought myself clothes; the kids always needed something.), but way more than that. Like time. Children require a huge amount of it. We're not just talking your basic feed 'em, keep 'em in clean clothes, but also entertaining them and even educating them. You can't pour into offspring without taking hours for playing with them, reading to them, talking with them, and driving them places. If you're not sure you're willing to sacrifice life as you know it, then think some more about whether you really want kids.

This isn't just on the part of the spouse with CF. Jon has always been a very involved parent. He put the kids to bed every night, helped with Cub Scouts or a soccer team, and did a good bit of the housework that he knew I wasn't up to doing. He never had time to go out with the guys or pursue his own interests.

You're also signing up for drama. If you like life to go smoothly with a minimum number of bumps in the road, parenting may not be for you. Every day something happens that's not according to plan. A beloved pet dies. A child's cut requires stitches. An entire gallon of milk spills. There are fights between your children, between them and their friends, even between you and them. Sometimes they're even in public and you're sure everyone in the world thinks that you're the world's worst parent.

That kind of stress is definitely not good for your health. So, if raising kids is important to you, make sure you have good stress relief techniques. A burned-out or depressed parent isn't a good thing, for either a child or his mom or dad.

Having a child means you have to have the "wisdom of Solomon". How else will you know which child broke

your antique plate when they both deny any involvement? What about when your teen wants to take the car out of state? Are you disciplining too harshly or not firmly enough? That's why it's important to have friends and/or family members whose opinions you trust. It's easier to stand strong in a decision if you have others who think that you're doing just the right thing. (A lot of prayer helps too!)

What's your energy level like? Kids not only have tons of energy; they suck every last ounce of yours. That means you have to have a plan to preserve yours. So take a nap when your baby sleeps. Get your kids in the habit of taking an hour after lunch in their rooms, where they can play quietly, read or whatever. Limit their activities to one a day so you won't be exhausted running them everywhere. And work out a deal with your teen, like we did, where he promised to make late nights (when he returned home after midnight) happen only on the weekends, so I can get the sleep I need, at least during the week.

Make sure your marriage is strong. A person married to someone with CF is usually pretty special and willing to deal with a lot of stuff your average spouse doesn't even have to consider. Parenthood takes even more. As I've mentioned, Jon has always done more than your "average" dad. And he's been willing to do it all without resentment. Before kids, make sure your spouse is totally on board with this, not just willing to go along with it because you want it.

Finally, think about what would happen if you died. Jon and I had some heavy discussions about this. Back when we got our kids, it was likely I would not survive to see them all grow up. Before our first adoption, he decided he was willing to commit to raising our children by himself if that happened. Thankfully, I'm still here, but there were several times when I was really sick and one or more of the kids asked Jon if I was dying. (He told them he didn't know.)

Yeah, it's true that parenting majorly changes your life. For us, it was worth it. But it's not for everyone. It's easy to fall in love with a cute little baby and think there's nothing you'd rather do than have one of your own. But those darlings grow up, and often at 2 or 10 or 17, they're not nearly so appealing. (Actually, they can be totally unappealing at times!) So if you're considering parenting, think it through very carefully and be sure you want and can handle it for all the years it takes. (That may mean the rest of your life—my 30-year old recently informed me that she's not ready for me to die yet!) ▲

Anne is 57 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Information from the Internet...

Compiled By Laura Tillman

This issue brings a potpourri of articles from the Internet

NEWS RELEASE

Polyphor Discovers A New Class Of Antibiotics With A Novel Mode Of Action

Polyphor Ltd published the discovery of a new class of antibiotics with a novel mode of action (Science, VOL 327, ISSUE 5968). This proprietary new class of antibiotics is effective against multi-drug resistant Gram-negative bacteria, opening up new treatment options for serious and often life-threatening infections. The most advanced drug candidate in this new class, POL7080, selectively kills the dangerous bacteria Pseudomonas aeruginosa. Polyphor is currently preparing the start of Phase I clinical trials with POL7080 to rapidly advance the clinical development and has initiated out-licensing negotiations with Pharma partners.

http://www.medicalnewstoday.com/articles/179924.php

Mpex Pharmaceuticals Presents Positive Phase 2 Clinical Trial Results of Aeroquin(TM) (MP-376) Treatment in Cystic Fibrosis Patients

Mpex Pharmaceuticals, Inc. announced the presentation of data from its Phase 2b clinical trial with Aeroquin™ (a proprietary aerosol formulation of levofloxacin, MP-376) in cystic fibrosis (CF). Trial results demonstrated statistically significant improvements in bacterial load, respiratory function and time needed for anti-pseudomonal antibiotics (a measure of exacerbations) versus placebo in a heavily treated patient population. To test the efficacy and safety on the new antibiotic called MP-376, a levofloxacin inhalation solution, the researchers recruited 151 CF patients and randomized them to receive MP-376 at three different dosing levels [120mg once a day (QD); 240 mg once a day, or 240mg twice a day (BID)] or

Continued on page 32

A Mother's Musings

By A. Nonymous

Being a mother with CF is … wait; can you even believe that this is a topic of discussion?! Twenty years ago the doctors would have told me that I would probably *die* as a child – forget *having a child!* It truly is amazing how far we've come!

In my mind's eye I see myself as a child, always playing mommy and baby with my dolls. As I entered the teenage years, I would turn to my younger cousins for maternal fulfillment. I read them stories, sang songs with them, and took them out for pizza every so often. I guess you could say that I always had a special place in my heart and in my life for kids. With this in mind, I thought that I would sail through the oceans of motherhood with ease and agility. Boy, was I in for a surprise!

Growing up in an Orthodox Jewish home, it was never a question of whether or not I would get married and have children…it was a given. When I was dating my soon-to-be husband and had that fateful conversation with him about my CF, one of his first questions was, "Will you be able to have children?". At the time I really had no idea what a loaded question that was, and my answer was a resounding, "Of course! Just because my lungs are not perfect doesn't mean my entire body is damaged!"

And so have children I did. I currently have two yummy, squishable, delicious guys ages 1½ and 5, and they are the light of my life. I did not do this without hesitation or careful planning. I would not jump on the pregnancy bandwagon without first discussing the situation with my CF doctor and meeting her criteria for healthy weight and PFT function, as well as careful consideration of the benefits and possible risks of pregnancy and subsequent child rearing.

Many people stop at pregnancy risks when considering children without realizing that raising children, especially for CF parents, can be even more physically challenging than birthing them. Sleepless nights abound, whether a baby needs to be changed and fed, a toddler has nightmares or a teen stays out till all hours of the night. And who has time to take proper care of herself when she has children who need her on a constant basis? (Case in point: there is no way that I could have finished this article if my husband hadn't stepped up and taken the kids out for a few hours!) I feel that pregnancy and childrearing for CF patients in general should come with one of those warning labels: "Enter at your own risk!" And after all is said and done, who's to say that my health would not have been better had I opted to forgo having children?

Even though I have made the decision to bring children into this world, I have to question every so often whether or not I made the right choice. My kids have a mother who clearly can't be Martha Stewart and Mr. Rogers all rolled into one perfect package. I am too busy taking care of my medication and PT regimen, fitting in exercise and keeping my meals healthy and hearty, to have gourmet suppers on the table each night and to run to the park every time the sun shines brightly. Guilt, therefore, plays a big role in parenting (for me). But I try to keep in mind what one of my dear friends reminds me when the necessity presents: If you raise your children with basic physical necessities, love, understanding, support, and morals (and a lot of prayer!) then they are already better off than two-thirds of the population. In addition, there are plenty of parents out there who don't do what I feel guilty about not doing, without having the excuse of having CF! How many children are picked up from school, fed, clothed, and at times even put to sleep by the babysitter?!

Although having children while continuing to care for myself at times seems daunting, even impossible, the times when it is managed with aplomb are simply indescribable. It is rewarding and enriching to have to care for others with love, patience, and devotion, when I take all of these things from others on a daily basis. I have helped create two special souls that can take the world to a better place with their smiles, hugs, laughs, innocent questions x 100/minute, and satisfaction at life's simplest pleasures (Mommy, guess what? This bug really lights up…cool…but he's so small – where is his battery?!). When everyone else sees me as less than average or ill, all I have to do is look into my guys' eyes and the fact that I am special and wonderful is reflected a thousandfold. Having kids also makes me a better person because I know that whatever I say or do, there are impressionable eyes and ears in every corner taking it all in.

The decision of whether or not to have kids while dealing with an all-encompassing illness such as CF is complicated, with many facts both emotional and physical that must be considered. For me, though, the quality of my life cannot be compared to what it would be like without my guys. I stand by my decision and hope that I will be a source of inspiration and strength for my little dudes for many years to come. ▲

The author has CF and lives with her husband and two sons in New York. She can be contacted at: A.Nonymous @yahoo.com

STENZEL BYRNES *continued from page 7*

CF has reminded me that my time is limited. With Emily and people like her, I believe having CF made her be the best person she could be. But CF *is* horrible; my friends and I have struggled, suffered, and endured pain. We worried about dying young and some of them, like Emily, did.

I think having CF reminds me that this disease is just one small part of greater human suffering. No one deserves their struggle; it's just part of being alive. Kriss and I have both asked at some point, "Why me? Why did I get stuck with CF?" The real question is, 'Why not me?' All humans suffer, though some suffer more than others. I just heard of a lady whose husband and son died within one day of each other. Life is so incredibly hard for so many.

Kriss feels that, "The answer to asking ourselves, "Why me?" is to accept that there is no answer. This is a hard thing to accept. Maybe bad things happen to good people for no more reason than chance or luck. Maybe, we need to try not to focus on why, but on *what* we are going to do with our individual challenges. And what kind of lifetime we are going to contribute to society and impact it with."

I find solace knowing that, together, Kriss and I have pondered why bad things happen to good people with CF. When people we love lose their battle with CF, it surely helps to know there are families and friends out there who struggle together to understand why. We hope you can join us in making spiritual sense of our shared CF struggles. ▲

Isa is 38 and has CF. She lives in Redwood City, CA. You may contact her at: isabel@usacfa.org.

WITONSKY *continued from page 15*

some reason, the light and warmth of the sun heals my aches and pains.

3. I really try not to do too much during the day, so I can conserve my full energy for when my girls come through the door at 4:15 PM and look forward to an energized Mommy. I fit an hour or so nap into my day. It took me a while not to feel guilty for taking a nap when I could have done X, Y and Z!

4. I try to have supper prepared before the kids get home, so I can have quality-time with my girls during the few hours between when they come home from school and until they go to bed.

5. My husband and I try to alternate the kids between doing homework and making sure that everything that needs to be prepared for school for the next day like snacks, lunches and, most importantly, clothing - literally from head to toe, shoes included – are ready so we are set up for an easy morning and start of the day.

6. The rest of the time, till 9:00 PM, is family fun time!

7. 9:30 PM sharp is bed time for both girls, and if they are in bed ready for the next day, I give each one special quality-time of 10 minutes each. Examples that they choose vary from playing a game to reading a book, tickle scratch, oil massage and their most favorite one - hearing me tell them stories of me when I was a young girl!

8. After my kids go to sleep, then it's my special time with my devoted husband, Yoni.

Understand that these are the basics that I do when I am feeling well. On days that I need help, I ask for it and people are very willing to help. Our day will still be okay, maybe not exactly the way I would have wanted it to be, but that's when my kids come to appreciate me for all the times I bend backwards and forward and go way over 200% of what other Mommies do!! ▲

Bracha is 33 and has CF. She and her husband, Yonason, live with their two healthy children in Brooklyn, NY.

USACFA Wants You!

Have you ever thought that you might like to be a director of United States Adult Cystic Fibrosis Assn. (USACFA)? Directors must have CF, must be at least 18 years of age and must live in the USA.

All directors must be willing to dedicate a few hours a month to USACFA business. Directors are expected to be active participants in USACFA, and to work on at least one subcommittee (e.g., Fundraising, Nominating, Editing, or History). Newly elected directors serve one-year terms, while re-elected directors serve two-year terms.

USACFA holds elections of directors every year in the spring, so let us know if you are interested. Send your résumé, no more than one-page please, to: cfroundtable@usacfa.org.

WELLNESS

Parenting With Cystic Fibrosis: What Was I Thinking?

By Julie Desch

I Was Warned.

But I like to think in terms of cost/benefit ratios. This is not a terribly romantic view of the world, but it has worked for me so far.

Here is what I was told: You are crazy to even think about this. Think of the germs! The colds, flu and who-knows-what-else you'll be exposed to! What will happen when you are sick? Who will care for them? Parenting is hard work! What about your job? What if your health suffers? You can't exactly return them!

Mind you, I was impressionable at thirty-something years old. It mattered to me what others thought, and I did listen to their arguments. Today, it isn't as unusual for a young woman with CF to become a mother, but this was fourteen years ago, and becoming a mom was much less common for someone in my shoes.

First, I was wearing Birkenstocks and I didn't care much about what was or wasn't common. I was stubborn and certain that only I could possibly decide what was right for my life. Second, I wasn't intending to go through pregnancy, labor or delivery. Instead, I was in the favorable (for me) position of having a female partner who wanted to be the birth mom. She didn't have CF or any other genetic illness, so this worked perfectly for us. She delivered and I adopted.

The Decision

I'm sure you are familiar with the term "informed" decision. This is a nice concept, but unless you have done it before, there is no way to have an honest clue about what you are getting into when you bring that newborn baby home with you. I have pictures of myself holding my oldest son for the first time, and there is honestly a look of sheer terror in my eyes. For good reason, it turned out.

The terror had nothing to do with me, or my health. It was simply the terror of a first time parent. So, yes, go through the pros and the cons. Think seriously about the future, and plan what will happen if you need to take time off for health reasons. You have to incorporate this into your decision-making process, unless you just want to wing it and hope for the best (not advised).

> *"When you fall head over heels in love with your baby, you suddenly understand why people do the strange things they do for their kids."*

JULIE DESCH, MD

But don't kid yourself and say, as I did, "I know exactly what I am doing!" What a joke.

Here is my advice: Listen to people...they love you. I'm sure they have your interests at heart. Certainly, listen to your doctors. They have no ulterior motive other than to keep you healthy. But don't forget to also listen to your heart, as only you can.

Planning

When I hear someone say that their pregnancy wasn't planned, that it just happened, or when I hear, "I was a mistake," I am simply amazed by how different experiences can be from one family to the next. Can I just say, we had to plan a bit? There was no luxury of a mistaken pregnancy in my life. It is not an easy task, mind you. We thought about timing, talked to potential donors, browsed through catalog after catalog of donor profiles, figured out who should be the stay-at-home Mom, logged early morning temperatures, ran to the doctor's office at just the right second with our precious and costly specimen, and then sat by the phone a couple of weeks later...waiting and wondering, anxiously.

Pregnancy

Fortunately, or not, I didn't go through this part. But I have coached people with CF who have gone through a pregnancy, so I'm not completely clueless.

You have to eat! You have to eat a ton. You have to eat more than you ever thought you could eat. And then you have to do it again the next day.

You also have to be closely monitored by your pulmonary doctors as well as your OB, because pregnancy with CF is not a low risk endeavor. You may also have blood sugar issues, so that will need to be monitored. Airway clearance will be a whole new animal, as your vest becomes too small, and your diaphragm starts to push up against your lungs. Continuing to move and stay as fit as possible through the entire pregnancy will make things much easier in labor and delivery, as well as with recovery and getting back on your feet when it's all over.

Infancy

Once the terror wore off, I was absolutely in love. When you fall head over heels in love with your baby, you suddenly understand why people do the strange things they do for their kids. There really aren't adequate words to describe this feeling. It's kind of a combination of amazement, adoration, enchantment and fierce protection bordering on obsession. Sound good? It is priceless.

Cute and cuddly as they are, though, there are a few downsides. You may come to believe you will never sleep again. You might, as I did, watch entirely too many middle-of-the-night infomercials and start thinking about buying real estate with "no money down!" You might wonder how in the world you will ever find time to bathe again. It will be a challenge to find a moment, let alone three, to use the bathroom in privacy.

Toddler Years

It just gets better. Now, not only are you in love with them, but they love you back! They bring you things, kind of like a cat proudly leaving a dead rat on the doorstep. You get mud pies, and already chewed food, and very creative drawings. You get to learn what a kid you still are—at heart, as you have more fun in the sand box than you really want to admit.

Ah, but there are some new challenges to deal with. What do you do with that poop in the tub? Will your child die first from Clorox poisoning, or will you kick off from E.coli pneumonia? Then there is the honest truth that this kid has no clue that you have needs. It doesn't matter how important they are. This takes some getting used to.

Preschool

I have two words to say, "Love notes." You will get many.

Preschool has many costs, though. First, you may literally spend a fortune during these years. It is not cheap. There are also some serious health concerns, and I'm being completely honest here. I was sicker than I had ever been during my first son's first year of preschool. Virus after virus, after virus, made its way to my mucosal membranes. Preschool kids don't understand the three-foot rule. They don't even abide by a three-centimeter rule. Be prepared mentally and psychologically. I don't know if it's possible to be prepared physically.

You also get a chance to answer many questions about your health. You'll be able to explain why you do treatments many, many times, not just to your own child, but also to random strangers on the street or in the grocery store after your child "outs" you. "Mommy wears a vest that shakes!" Inquisitive looks from the four people in front of you in line...

Of course, it's all worth it and more, when your child looks at you and adoringly declares, "You're Big! You're Strong! You're Mommy!"

Yes, he really did.

Elementary School Age

Good things happen during this time. Many, many good things. You get to volunteer hours and hours at school and still feel guilty when you say, "No." But in this process, you meet countless new friends, and get out and about in the world. I found that this activity, lots of it, really helped me to focus less on my health and my own worries, and more on having fun and being involved. You also may find, as I did, an entirely new and unique airway clearance technique in the form of hysterical laughter. This occurs with greater and greater frequency as your kid develops his or her sense of humor and brings home friends who are doing the same. Jokes get played on you. Things get thrown. Sometimes mysterious holes appear in your walls. The downside of laughing spasms is early onset incontinence; but worse things could happen, right?

It is also finally possible now to leave them long enough to go to the bathroom. A television or video game console is required, but it's such a relief, you won't care. You've been holding that bladder for a long time. Perhaps this might have something to do with the stress incontinence?

But the best part about these years is watching them grow. Watching proudly when they read "Cat in the Hat" alone for the first time (so what if they have it memorized?) is a moment to behold. Going to winter music concerts each year, you can observe how every year, the songs they sing sound increasingly more like real music that you recognize. And then, there's always the Mother's Day breakfast-in-bed fiasco, I mean surprise.

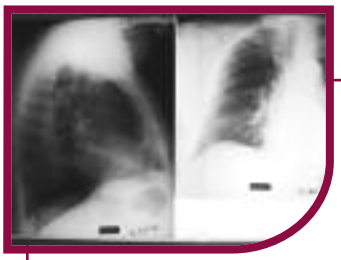
And to think, had I listened to "them", I could have missed all this?

Teenage Years

What was I thinking?

They were right...

Julie is 49 and is a physician who has CF. You may contact her at: jdesch@usacfa.org



A DEEP BREATH IN
Being Parents To Ourselves

By Debbie Ajini

This column is a bit of a stretch for this issue's Focus topic. Originally, this column was meant for the *Spring 2009* issue, which related to work issues. Accidentally, it was not published. So, I thought maybe I could somehow make it apply now! Although this is a stand-alone column, in that I do not need to follow the Focus topic, I tend to do so because I think it is great to have as many perspectives as possible. This column is about working and, basically, how I had to become my own parent and make some tough decisions for me and my health. As you may or may not have experienced, every adult with CF will have to evaluate his/her position in the working world at some point. We each have to ask our own unique questions as each person's situation is different.

In my situation, I had to decide if working where I was, was worth getting recurrent infections. As a teenager and young adult, I mainly worked in the childcare industry. I loved it. I enjoyed working with the kids, but I kept getting sick over and over. Looking back, I now know that working with kids was not the best choice for me, especially because I did not have the level of awareness about infection control that I do now.

I remember the moment I knew it was not worth the repeated infections. I was working in a daycare, and I had a 103 degree fever. I felt so bad that I lay on the floor next to the kids when they were having their nap time. I knew then that working wasn't good for me. I had worked for eight years before I got to this point and made the choice to apply for Social Security Disability Income (SSDI). I applied in early 1994 and after being rejected once and hiring a lawyer, I was approved in mid-1995.

In my situation, I was very fortunate in that I had a fiancé who was ready and willing to step up and provide for me. And since we were planning to be married, I would also have health insurance. I realize not everyone may have this cushion, but we still have to ask ourselves what our health is worth.

It is a very tough choice and a very individual choice whether to work or not. Everyone has his/her own beliefs and guidelines for what is doable. For me, I wonder if I had worked in another field besides childcare, could I have worked a few more years? Then, on the other hand, I think that maybe because I did quit working, I was able to stay pretty healthy from 1995-2005. I will never know.

> *"I wonder if I had worked in another field besides childcare, could I have worked a few more years?"*

DEBBIE AJINI

I do have my own business. I have had it for seven years now. It is nice to be able to make the extra money when I want to. At least I feel I can contribute to our household in some small way. In the first few years I was really excited about what I was doing, and my health was stable, so I did more. As I have had more issues in the past few years, I have been able to easily adjust my workload. Some years I work a lot, and last year I did not work at all. This year I am resuming work with only one of my clients. My goal is just 10 hours a month. It's not a lot, but with the way the economy is right now, it will help.

I do think about what it would be like to have a full-time job and bring home a 40-hour paycheck; the extra money would be so nice. But I know I would not be able to do anything else but work, eat, sleep and my treatments. And I can only imagine how quickly I would end up on IVs anyway.

It was tough to step into a more parental role (like how I tied that back in to the Focus topic?!) and make the decision. So, I am very lucky that I can choose not to work and put my health first. This is one of the many things I am grateful for every day. I only wish all adults with CF could make the healthiest choice for themselves without worrying about paying bills or having health insurance. Maybe someday that will be possible. ▲

Debbie is 39 and has CF. She is a Director of USACFA and is the Secretary. Her contact information is on page 2.

MEET THE NEW DIRECTORS

Meet Jen Eisenmann


JEN EISENMANN

I was born in the middle of a blizzard in New York City on New Year's Day, 1971. Diagnosed with CF at the age of one, I remained an only child. At age 2, my parents and I moved to Connecticut where I graduated from high school. I decided I wanted to be more independent and get away from the cold weather, so I applied to, and got accepted into, Furman University, a small college in South Carolina. In 1992, I graduated with a degree in Religion and Psychology.

After taking a three-month training course in Colorado, I became a youth minister at an Episcopal church outside of Lansing, Michigan, where I lived for six years. It was there, when I was in my early twenties, that CF really began to affect my life greatly and I started to take my care seriously.

In 1999, I moved back south to Aiken, South Carolina, and became an accounting assistant at Dogwood Stable, Inc. Unfortunately, I had to retire to take better care of my health in 2005. In an interesting twist of fate, since the company downsized last year, I now work there again, two afternoons a week, in the same position I previously held.

In 2001, I married my wonderful husband, Pete, who is a Leasing Agent with a popular local real estate company. Over the past year, he has taken up running, which he uses to raise money for CF. We live with our two crazy dogs, Cinnamon and Lily.

I have helped raise my amazing stepchildren, Jason, who was 14 in 2001, and Jessie, then 11. Now 24, Jason served in the Marines for four years (he was deployed to Iraq three times) and currently lives and works in Aiken. Jessie, now 21, is going to be a senior at the University of Georgia Honors College. She would like to serve in the Peace Corps for two years after she graduates.

I have a zany sense of humor and enjoy listening to music of all types, crocheting, and hanging out with my friends at the local brew pub. Pete and I are heavily involved with an annual CF fundraiser in Columbia, SC. ▲

See Jen's contact information on page 2.

Meet Anne Williman


ANNE WILLIMAN

I'd like to introduce myself to *CF Roundtable* readers. Having been a loyal subscriber of this publication for many years, I'm delighted that recently I have been elected to serve on the USACFA Board of Directors.

I was diagnosed with CF as a five-year-old. Back then, the sweat test consisted of putting the entire body, from the neck down, into a plastic bag and lying there for hours until enough sweat was collected. I remember my mother reading to me as I did this. Despite the fact that I had no lung symptoms at that time, the diagnosis came back CF, and my parents were told I would not live to see age ten.

Yet here I still am, and I thank God for the full life He's given me. I went to college (graduating with a BA in English and religion), married my husband Jon (we recently celebrated our 35th anniversary), and raised three children whom we adopted when they were infants. (They are now ages 30, 26, and 18.) I even have the most darling 18-month-old grandson!

Professionally, I have worked as a freelance writer since college. This has been the perfect career for me, as I write out of a home office and can adjust my schedule according to how I feel physically. I've been blessed with some success as I've sold two books (plus another I co-authored) and over 700 magazine articles. In addition, my husband and I own a vending machine route, and I handle the web site, pay the bills, invoice customers, and do whatever else needs to be done.

In my free time, I like to read, do crafts, and with my husband, walk Shekinah and Rhema, our two black Lab mixes. We also enjoy traveling, having taken trips to Brazil, England, Israel, Mexico, Canada, and Italy, as well as many parts of the US, including California, Texas, Florida, and Maine.

I have participated in numerous clinical trials, and I have the majorly scarred veins in my arms to prove it!

I look forward to doing all I can as a director for USACFA. ▲

See Anne's contact information on page 2.

UNPLUGGED...
With Akihiro Adachi

By Richard De Nagel

Hello everyone and, of course, welcome to our next edition of Unplugged. This interview is very powerful and will open your eyes to what CF is like in another part of the world. Our interviewee comes from Japan, thanks to Isa Stenzel Byrnes and Ana Stenzel. His name is Akihiro and they met him on their tour of Japan, last fall. They found his story so compelling that they asked that I interview him. After talking to them and him, I could not pass up his story. So here we go.

AKIHIRO ADACHI

After reading through his interview, I was speechless. I know I struggle with CF and I know that it is tough to deal with on a daily basis, but I take for granted all the resources, community and people involved with my disease. I am not going to be so presumptuous anymore. So, without further ado, I will let Akihiro tell his story.

1) What is your name, age and where do you live? Akihiro Adachi, 21 years old. (Born April 29, 1988) I live in Sendai City, Miyagi, Prefecture, Japan

2) With whom do you live? Or, who is in your family? I live with my father, my mom and our dog, Lucky. Family structure is the same.

3) When were you diagnosed with CF? How? My parents were told I had abdominal bloating during a fetal echo, and I was born on the 37th week through vacuum extraction. On the first day after I was born, I was diagnosed with a bowel blockage, and had surgery to remove the ileum. After the surgery, due to obstruction, cholestatic condition occurred. From my 3rd month I had continuous refractory diarrhea and bronchitis. When I was four months old, I was suspected of CF from the long lasting coughs and diarrhea. They checked the chloride concentration in sweat, with a result of a high figure of 96mEq/L. After this test, I was diagnosed with CF.

4) How is your health right now? I completed my IV treatment, so I am relatively stable.

5) What is your daily medical routine like? Do you do nebulizers, chest percussion, vest, IPV, IV antibiotics, inhaled antibiotics, enzymes, etc? How often? I use the nebulizer and IPV four times a day. I used IV antibiotics ten times the year before last, and five times last year. I take inhaled antibiotics three to four times a day. For enzymes, I take six grams of Creon10 per day.

As far as chest percussion is concerned, it has been difficult, so I am hoping to use a vest. There are only a few in Japan, so most hospitals don't have them. In addition, using the vest at home cannot be applied for insurance, so I cannot use it as I wish.

6) Are you compliant? Do you do all your treatments like your doctor wants you to? Yes, I am compliant. I do all the treatments my doctor tells me to.

7) Where do you receive your medical care? Is your doctor a pediatrician, pulmonologist, or another specialist? I receive my medical care at the hospital at Tohoku University Hospital and at home. I have two doctors: one is a pediatrician who overlooks my nutrition and overall situation, and the other is a pulmonologist who checks on my lung situation.

8) Does your Japanese doctor treat other CF patients? Also, have you ever received care from a CF specialist (like in the US, Denmark; when)? I am the only CF patient for my doctor.

In 1994 (when I was six), and in 1999 (when I was 11), I went to the Los Angeles Children's Hospital. From 2003-2006, I was treated at Rigshospitalet (Denmark) once a year. Since 2007, my father visits Denmark once a year to consult.

9) How often do you go into the hospital? What is it like? I was in the hospital for four years, from right after I was born until I was four years old. During the four years, I had high-calorie nutrition 24 hours a day from my central vein. After that, I would be in the hospital when a bad infection

occurred - once or twice a year.

10) How often do you use home IV antibiotics? In 2008 I had IV antibiotic treatments ten times for a course of two to three weeks. Out of the ten, three were while I was in the hospital. In 2009, I used home IV antibiotics five times.

11) What are your thoughts about having CF? I am working hard on my treatment, as the representative of CF patients in Japan. I believe the result of my efforts will help the future Japanese children with CF.

12) Do you think CF is a good or bad thing? Please explain. I first thought CF was a bad thing that torments me. Now, I feel like it "is what it is" that I was born with CF, and I don't expect anything.

13) What do you do for fun? I take a walk with my dog, Lucky.

14) What are you interested in? Watching sports such as baseball and soccer.

15) What type of music do you listen to? Beatles

16) What's your favorite TV show and movie? Do you watch American TV? Apollo13, Angels in the Outfield, Back to the Future, and Hachiko: A dog's Story (with Richard Gere)

17) How do you get through the bad days (when you are sick, when you can't breathe, when you haven't left the house all day, when you feel hopeless)? For a change, I would watch my favorite comedy shows, or the Olympics, or baseball and soccer games.

18) Are you in a relationship? Do you want to be? No, I am not in a relationship.

19) Do you have friends who know about your CF? No, I don't have friends who know I have CF.

20) How have your parents handled your CF when you were younger, and now as an adult? Work hard (ganbaru) and work together as a family.

21) Do you believe in God, or a Higher Power? Can you explain how your spiritual beliefs impact how you deal with CF? I wish that there were angels as in the movie "Angels in the Outfield."

22) What do you look forward to most in the future? I would like to go on a trip abroad.

Every time I read his story I am struck by the simplicity, isolation and strength of this young man. Imagine having to be the poster child for all people with CF in the US. I have to sit down at that thought. Could you imagine you were the only CF patient your doctor saw? CF is a small disease but, once you get out of the US or Europe, it gets so much smaller and there are fewer resources, doctors and financial assistance. Gratitude is all I can think of at the moment.

I have another hero to add to my list. His foresight to see that he needs to help future generations is amazing, especially since he is one of the only adults there now. I am not even getting to all the cultural aspects of Japan that limit him. Mainly, they do not speak about having any ailments and it is frowned upon to talk about it. So his desire to be an advocate for others makes him that much more remarkable.

I will end there and leave you with the thought that I am going to say, "thank you," to everyone involved with my CF care. I want to see how many people I can thank in a day or so. Try it. Enjoy!! ▲

Rich is 41 and has CF. He lives in San Francisco, CA and may be contacted at: rdenagel@usacfa.org.

IMPORTANT CHANGES

You may have noticed that USACFA and *CF Roundtable* have experienced some changes. There are new officers and board members in our organization. *CF Roundtable* has a different column: "Editor's Notes" rather than "A Word from the President." Also, USACFA has expanded to the point where we have two separate addresses for different purposes. We ask that all correspondence that does not include any money in it be sent to the old PO Box number and that all mail containing money of any kind be sent to the new PO Box.

When sending in a subscription or donation of any amount, send it to: **USACFA, PO BOX 151024, ALEXANDRIA, VA 22315-1024.**

All articles, general inquiries, comments, questions, or praise should be sent to: **USACFA, PO BOX 1618, GRESHAM, OR 97030-0519.**

TILLMAN *continued from page 23*

placebo. On average, the patients were 29 years old, had received almost five courses of inhaled antibiotics active against P. aeruginosa in the past year, were on multiple treatments to control their CF lung disease, and had just slightly more than 50 percent of predicted lung function for healthy individuals. After 28 days of treatment, patients receiving MP-376 had reduced P. aeruginosa density in their sputum compared to those who had received placebo, and showed no evidence of antimicrobial resistance. The greatest significant treatment effect observed was in the patients who received the highest dose of MP-376 of 240mg twice a day. Lung function also improved in all the MP-376 groups with the greatest improvement being seen in those who received the 240mg BID dose, with an average improvement in FEV_1% predicted of 10.9 percent over placebo. A similar effect was also seen in other measures of lung function.

Treatment with MP-376 reduced the need for treatment with other anti-pseudomonal antimicrobials, with a 79 percent reduction over placebo in the MP-376 240mg BID group. While the reduction in P. aeruginosa density was expected, the degree of lung function improvement and reduction in the need for other anti-pseudomonal antimicrobials in this heavily treated patient population was unanticipated. Phase 3 trials are scheduled to begin later this year.

http://tinyurl.com/2fgzx5g OR http://www.newswise.com/articles/view/564407/?sc=mwhn OR http://www.mpexpharma.com/pr_20100517.html

Data Suggest Denufosol May Hold Promise as an Early Intervention Treatment for Cystic Fibrosis

The data suggest that denufosol, an inhaled ion channel regulator, has properties that allow it to reach and improve lung function in the small airways, which may support its potential as an early intervention therapy. In vitro data suggests that denufosol, as a result of its aerosol and chemical characteristics, has the potential to reach the small airways of the lungs, where CF lung disease begins. The potential of denufosol to reach the lung's small airways was supported by a post-hoc analysis of the TIGER-1 trial, which showed a significant improvement relative to placebo in FEF25%-75% (Forced Expiratory Flow) in a subgroup of patients (n=329) with baseline FEV_1 (Forced Expiratory Volume in One Second) less-than or equal to 110% predicted normal. In the TIGER-1 trial, following inhalation, there was little to no evidence of systemic exposure to denufosol and no evidence of accumulation as measured by plasma levels. In the trial, denufosol was well-tolerated and had a favorable safety profile. Denufosol is a first-in-class ion channel regulator targeted as an early intervention treatment that potentially corrects the ion transport defect in patients with all CF genotypes. Denufosol is designed to enhance airway hydration and mucociliary clearance by increasing chloride secretion, inhibiting sodium absorption and increasing ciliary beat frequency. These integrated pharmacological actions and the potential to reach the small airways are key to maintaining lung function and potentially delaying the progression of lung disease. Inspire is currently conducting its second Phase 3 clinical trial with denufosol, TIGER-2.

http://tinyurl.com/b2t3p5t

Orphan Drug Research Offers Hope

In addition to their suffering, rare disease patients often have to face the harsh reality that few pharmaceutical companies will ever be able to offer new treatments for their condition because the costs of new treatments will never be recovered from such a small market. But there are ways they can be helped. The U.S. Food and Drug Administration's "Orphan Drug Designation" offers a wide range of benefits that help organizations developing treatments for diseases and conditions affecting fewer than 200,000 patients in the United States. It was recently granted to McGill University for research conducted at the MUHC into the use of the drug fenretinide for the treatment of pulmonary infections caused by Pseudomonas aeruginosa in patients with Cystic Fibrosis (CF). Fenretinide reduced lung inflammation as well as the frequency and the severity of pulmonary infections.

http://www.medicalnewstoday.com/articles/190654.php

Centocor Ortho Biotech Inc. Acquires RespiVert Ltd., Strengthens Pulmonary Focus

Centocor Ortho Biotech Inc. announced that it has acquired RespiVert Ltd., a privately held drug discovery company focused on developing small-molecule, inhaled therapies for the treatment of pulmonary diseases. The company's lead compounds, RV-568 and RV-1088, narrow spectrum kinase inhibitors with a unique profile of anti-inflammatory activities, are progressing into clinical development as potential first-in-class treatments for moderate to severe asthma, Chronic Obstructive Pulmonary Disease (COPD) and Cystic Fibrosis (CF). The clinical development of RV-568 and RV-1088 will be led by RespiVert in collaboration with scientists at Centocor Research and Development, Inc. The RespiVert compounds offer the potential for a new class of medicines for patients with severe lung disease who are insensitive to inhaled corticosteroids.

http://www.medicalnewstoday.com/articles/190810.php

FYI

Immune-Mediated Severe Hemolytic Crisis with a Hemoglobin Level of 1.6 g/dl Caused by Anti-Piperacillin Antibodies in a Patient with Cystic Fibrosis. S. Kunzmann, W. Thomas, B. Mayer, S. Kuhn and H. Hebestreit

Infection. Published online: March 5, 2010

A 23-year-old female patient with cystic fibrosis developed severe intravascular hemolysis with a minimal hemoglobin level of 1.6 g/dl after 7 days of treatment with piperacillin, consistent with an immune-mediated hemolytic crisis. Twenty days later, the patient could leave the hospital in good condition without any neurological deficit. This is the lowest reported hemoglobin value caused by hemolytic anemia with intact survival. As piperacillin is commonly used in patients with cystic fibrosis, it is important to monitor the full-blood counts of patients during treatment with piperacillin and to be aware of the potential for hemolytic anemia to develop. Anti-piperacillin antibodies should be considered whenever these patients develop hemolytic anemia or a positive direct antiglobulin test (DAT). Furthermore, drug-fever under piperacillin application could be a warning sign for the development of hemolytic anemia.
http://tinyurl.com/ydz7jgy

A 1-m distance is not safe for children with cystic fibrosis at risk for cross-infection with Pseudomonas aeruginosa. Filippo Festini, RN, BA, BSN, Giovanni Taccetti, MD, Valeria Galici, MD, Stella Neri, RN, BSN, Sofia Bisogni, RN, BSN, MSN, Daniele Ciofi, RN, Cesare Braggion, MD. American Journal of Infection Control. Volume 38, Issue 3, Pages 244-245 (April 2010)

Although maintaining a distance of 1 m between persons with cystic fibrosis (CF) is a universal recommendation to prevent respiratory cross-infections such as Pseudomonas aeruginosa, evidence supporting this preventive measure is scarce. Examining 336 samples from 42 patients with CF collected experimentally from sterile surfaces after speaking and coughing, we found that transmission of P aeruginosa beyond 1 m is possible during both talking and coughing, although the probability is low (1.7%).
http://tinyurl.com/y8qbbwu

Cystic fibrosis and survival to 40 years: a case-control study. N.J. Simmonds, S.J. MacNeill, P. Cullinan, M.E. Hodson. Eur Respir J 2010

Factors resulting in increased probabilities of survival included: high BMI (Body Mass Index), FEV_1 and FVC at transfer to the adult clinic and the exclusive use of oral antibiotics. Factors resulting in decreased probabilities of survival: P. aeruginosa acquisition or pneumothorax before transfer to the adult clinic and referral from a paediatric clinic in a deprived area. Long-term survival is associated with the clinical features present by the time of referral to an adult clinic; even "early-diagnosis" disease appears to have different phenotypes, possibly independent of CF gene function, that have different survival patterns.
http://tinyurl.com/y4pw7ph

Lack of correlation between pulmonary disease and cystic fibrosis transmembrane conductance regulator dysfunction in cystic fibrosis: a case report. Hara Levy, Carolyn L. Cannon, Daniel Asher, Christopher Garcia, Robert H. Cleveland, Gerald B. Pier, Michael R. Knowles and Andrew A. Colin. Journal of Medical Case Reports 2010, 4:117

Despite a prevailing idea in cystic fibrosis research that the amount of functional cystic fibrosis transmembrane conductance regulator predicts clinical status, the results indicated that respiratory disease severity in cystic fibrosis exhibits phenotypic heterogeneity. If this heterogeneity is, in part, genetic, it is most likely derived from genes outside the cystic fibrosis transmembrane conductance regulator locus.
http://tinyurl.com/2dz6gqv

Failure to Recover to Baseline Pulmonary Function after Cystic Fibrosis Pulmonary Exacerbation. Don B. Sanders, Rachel C.L. Bittner, Margaret Rosenfeld, Lucas R. Hoffman, Gregory J. Redding, and Christopher H. Goss. Am. J. Respir. Crit. Care Med. 2010

Patients with cystic fibrosis periodically experience pulmonary exacerbations. Previous studies have noted that some patients' lung function (FEV_1) does not improve with treatment. Of 8,479 pulmonary exacerbations, 25% failed to recover to baseline FEV_1. A higher risk of failing to recover to baseline was associated with: female gender; pancreatic insufficiency; being undernourished; Medicaid insurance; persistent infection with P. aeruginosa, B. cepacia complex, or MRSA; ABPA; a longer time since baseline spirometric assessment; and a larger drop in FEV_1 from baseline to treatment initiation.
http://tinyurl.com/26swnlo

REPRODUCTION

Cystic Fibrosis in Pregnancy. Whitty, Janice E., MD. Clinical Obstetrics and Gynecology: June 2010 - Volume 53 - Issue 2 - pp 369-376

Women with cystic fibrosis (CF) are living to childbearing age and many have successful pregnancies. Preconception care with optimization of pulmonary function, eradication of pulmonary infection, improved nutritional status, and diabetes care improve fertility and pregnancy outcome. Women with CF, poor pulmonary function and nutrition, and less than ideal body weight are more likely to suffer adverse outcomes. Women with CF and pulmonary hypertension risk mortality. Individuals with CF and end stage lung disease have improved survival after lung transplant. Women with lung transplants can have successful pregnancies, but the risk of organ rejection and death are high.
http://tinyurl.com/232yszq

Pregnancy and Cystic Fibrosis.

Continued on page 34

Edmund M.T. Lau, Carmel Moriarty, Robert Ogle, Peter T. Bye. Pediatric Respiratory Reviews. Volume 11, Issue 2, Pages 90-94 (June 2010)

The management of a pregnancy in a woman with cystic fibrosis is usually achieved with successful outcomes for mother and child with appropriate multidisciplinary care. The process begins prior to conception and requires frequent monitoring of the mother's respiratory status, level of glycemic control and obstetric wellbeing. Recent reports have suggested that pregnancy can be managed without a persisting decrement in lung function beyond what may be expected in women with cystic fibrosis who are not pregnant. With the increasingly positive outcomes for people with cystic fibrosis, it is likely that more couples will choose to pursue pregnancy, cognizant of the risks and longer term issues for mother, child and family.
http://tinyurl.com/24mhcmy

Fertility in Men with Cystic Fibrosis Assessment, investigations and management. Howard C. Smith. Pediatric Respiratory Reviews. Volume 11, Issue 2, Pages 80-83 (June 2010)

Congenital absence of the vas deferens is the primary cause of azoospermia in men with cystic fibrosis [CF]. Sperm capable of fertilizing mature oocytes in vitro can be extracted from the majority of men with cystic fibrosis. Mature spermatozoa can be obtained by a simple percutaneous needle aspiration of the epididymis or testis after local anaesthetic has been infiltrated around the spermatic chord. This procedure has been successfully repeated up to six times in some men. Epididymal or testicular sperm can be used to fertilize mature oocytes by intracytoplasmic injection in vitro. All offspring resulting from such procedures will be CF carriers and to reduce the probability this will result in a child with CF it is important the female partner has pre-treatment screening for CF mutations.
http://tinyurl.com/2g3woy2

Contraception, Communication and Counseling for Sexuality and Reproductive Health in Adolescents and Young Adults with CF. Anna Tsang, Carmel Moriarty, Susan Towns. Pediatric Respiratory Reviews. Volume 11, Issue 2, Pages 84-89 (June 2010)

With survival now into the fourth decade and rapid growth of the adolescent and adult population of people with cystic fibrosis CF sexual and reproductive health issues are integral to the management of adolescents and adults with CF. Education and counseling for sexual health related issues must be included in the daily routine of CF care. With advances in genetic counseling, contraception, assisted reproductive technology and collaborative management, adolescents and young adults with CF realizing their sexual and reproductive potentials safely and realistically can be possible .
http://tinyurl.com/24azxoe

BACTERIA AND FUNGI

Isolation of the fungus Geosmithia argillacea in the sputum of people with cystic fibrosis. R. C. Barton, A. M. Borman, E. M. Johnson, J Houbraken, R. P. Hobson, M Denton, S. P. Conway, K. G. Brownlee, D. Peckham, and T. W.R. Lee. J. Clin. Microbiol. doi:10.1128/JCM.00184-10

The repeated isolation of the fungus Geosmithia argillacea from the sputum samples of people with cystic fibrosis is reported. Identification was based on morphology and DNA sequence analysis. Isolation of G. argillacea did not appear to be associated with clinical deterioration.
http://tinyurl.com/24925s u

Sputum Candidia Albicans Presages FEV_1 Decline and Hospitalized Exacerbations In Cystic Fibrosis. Sanjay H. Chotirmall, MD, Elaine O'Donoghue, MD, Kathleen Bennett, PhD, Cedric Gunaratnam, MD, Shane J. O'Neill, MD and Noel G. McElvaney, MD. Chest. Published online before print May 14, 2010

The role of Candida albicans in the cystic fibrosis airway is underexplored. Considered a colonizer, few question its pathogenic potential despite high isolation frequencies from sputum culture. Independent associations of colonization with clinical outcomes were determined and the longitudinal effects of C. albicans acquisition on body mass index (BMI) and forced expiratory volume in the first second (FEV_1) evaluated. Colonization with C. albicans was frequent and best predicted by pancreatic insufficiency, osteopenia and co-colonization with Pseudomonas spp.. C. albicans colonization significantly predicted hospital-treated exacerbations after adjustment for confounders. Exacerbation rate significantly increased in the chronically or intermittently colonized following first acquisition of C. albicans. Colonization accelerated rates of decline for BMI and FEV_1.
http://tinyurl.com/2ufo7o2 ▲

Laura Tillman is 62 and has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.

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- You can reach **USACFA** and ***CF Roundtable*** at anytime by phone or fax at **(503)669-3561**. (That number always answers by machine.) You may email us at **cfroundtable@usacfa.org**
- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq., 811 Rusk Street, Suite 712, Houston, TX 77002-2807**.

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

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KATHY RUSSELL 5/11
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IMPORTANT RESOURCES

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

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

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

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

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