CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS SUMMER 2009

For Better, For Worse:

His Story

By Steve Downey

Of all the dog runs in all the towns in all the world, she walked into mine, and was I lucky she did. This cutie-pie with the Boston Terrier seemed to like me. We chatted, then hobnobbed; it progressed to flirting and pretty soon, we had a date. She asked if I liked sushi. I didn't. I thought eating uncooked fish was something humanity should have left behind with stone tools and human sacrifice. "I would love to try some sushi with you," I said. This girl was worth the risk.

She let me know about her illness on our date. "I had a double lung transplant," she said. "Why?" I asked. "I have CF." I knew what those initials stood for, but that was the sum of my knowledge. Cystic fibrosis was something there was the occasional article about in "The New York Times", and wasn't there a telethon for it? She explained it to me then, on our date. Chloride. Sticky mucus. Genetic. I wasn't paying attention - too busy trying to get the raw fish to pass down my throat. And I was more interested in pursuing this

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Her Story

By Andrea Eisenman

Most people with cystic fibrosis (CF) know that you can catch something from other people with CF, be it B. cepacia or other organisms carried in their mucus—cross-infection. And, of course, we know we can catch a cold or sinus infection from anyone, CF or not. But I did not realize that it was possible to keep getting sick from my newly married spouse.

Since Steve and I have been together, I started to notice that I was getting more sinus infections than usual. Normal for me was 2-3 sinus infections a year and I usually never got sick during the cold season. That changed as well. I started thinking, could I be getting this from Steve, my husband? We do have an agreement that the minute he starts to feel a cold come on he has to tell me, and I do the same. But he sometimes sounded congested and sneezed at night. I would ask him if he was sick and he would say, "No." I then felt stupid asking, "Do you have a sinus infection?" The answer was, "No." But he sounded to me like he

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STEVE DOWNEY AND ANDREA EISENMAN

INSIDE THIS ISSUE

CF ROUNDTABLE
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A WORD FROM THE PRESIDENT...

Dear Readers,

After four years as President of USACFA, I have decided to step down and allow another USACFA Director to assume the responsibilities. I have extremely enjoyed working with the Board of USACFA, and I hope to continue to contribute to this wonderful organization as a Director. **Kurt Robinson** has resigned and we are holding our elections as I write. In our next newsletter we will introduce the new board members and officers.

Health concerns are my primary reason for stepping down, as I have been battling my second bout of rejection since my transplant almost five years ago. I am stable for now, and I have been blessed to have made it this far post-transplant as, statistically, only about 50% make it this far. It continues to be a battle for adults with CF, even after transplant, to live that "normal" life; it is just as important to continue to hear *Transplant Talk* from others who have made this courageous decision.

On the front page, **Steve Downey** and **Andrea Eisenman** share one of their "for better or worse" experiences since getting married, yet another lesson for our supportive spouses! In *Ask the Attorney*, **Beth Sufian** concentrates on dealing with payments to both providers and collection agencies, and gives some sound advice on handling both. I read **Isa Stenzel Byrnes'** *Spirit Medicine* column and sent it directly to my father and step-mother, who had just recently moved – a move that took seven weeks, after 40 years in the same home. Read and you'll understand, and maybe take heart! **Rich DeNagel** interviews his good friend, **Steve Gonzalez,** in *Unplugged* and provides interesting background about their CF relationship and Steve's personal life.

In *Voices from the Roundtable*, **Rachel Mauger** prepares for transplant. Rachel's story, to her current age of 26, very much reflects my own, and I found her advice to be very sound. I had to do some fun math on a part of our *Milestone* section – the average birthday of our seven people was 45.5 years – that's just beautiful. **Debbie Ajini** attempts to describe the meaning of her column, *A Deep Breath In*, and how it might differ from the rest of our newsletter.

In *Transplant Talk*, **Colleen Adamson** tells the story of her unusual child, **Penny,** and the joy Penny brings to her life. That leads to our focus topic, "Becoming a Parent with CF". I took the liberty of discussing the adoption of my daughter, **Sarah,** and the opportunities and challenges which that brought to my life. **Susie Baldwin** provides us a viewpoint that I'm sure many other CF adults share – that it is just fine to be childless. **Cara Brahm** discusses having a child with CF and the challenges and enjoyment it has brought to her and her family. **Jeanie Hanley, Debra Radler, Lynn Williamson,** and **Anne Williman** also share their perspectives, as does **Kathy Russell** in her *Speeding Past 50* column. Enjoy all of these as you read through this issue!

Peace,

Publication of *CF Roundtable* is made possible by donations from our readers and grants from The Boomer Esiason Foundation, CF Services, and Genentech, Inc.

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

Victor Cellers
Broomall, PA
46 on March 8, 2009

Kathi Novelli Clapham
West Chester, PA
38 on June 25, 2009

Amanda Cokelet
Hazlet, NJ
30 on June 6, 2009

Paul Feld
Florissant, MO
52 on May 9, 2009

Kathy Harris
Bowie, MD
66 on May 17, 2009

Debra Radler
Roselle, IL
47 on May 31, 2009

Stephanie Rath
Brownsburg, IN
40 on January 2, 2009

Wedding

Victor & Deborah Cellers
Broomall, PA
16 years on January 9, 2009

Paul & Kristi Feld
Florissant, MO
18 years on June 1, 2009

Debra Radler & Adrian Gulinski
Roselle, IL
2 years on June 8, 2009

Kathy & Richard Harris
Bowie, MD
46 years on June 22, 2009

Arthur & Brandie Herron
Carmichael, CA
1 year on June 21, 2009

Stephanie & Randy Rath
Brownsburg, IN
12 years on May 3, 2009

Transplant

Kathi Novelli Clapham, 37
West Chester, PA
Bilateral lungs
3 years on May 29, 2009

Amanda Cokelet, 30
Hazlet, NJ
Bilateral lungs
10 years on June 16, 2009

Ray Corwin, 47
Jacksonville, FL
Bilateral lungs
12 years on April 26-27, 2009

Kathy Harris, 66
Bowie, MD
Bilateral lungs
3 years on July 10, 2009

NEW BEGINNINGS

Transplant

William H. Coon, Jr., 49
Spring, TX
Bilateral lungs
On January 23, 2009

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) 2009: Becoming A Parent With CF.
Autumn (November) 2009: Gender-related Problems in CF. (Submissions due September 15, 2009) There are many problems with having CF that can be directly related to one's gender. Do you have any questions about or experience with any of these issues? Please share with our readers.
Winter (February) 2010: Diet and Nutrition. (Submissions due December 15, 2009) When you have CF, keeping a healthy body can be a trial. Do you have experience with maintaining an ideal weight and body mass? Do you have good ideas for ways to cook and/or eat a healthier diet? Please share your information with us.
Spring (May) 2010: Traveling For Work or Pleasure With CF. (Submissions due March 15, 2010)

ASK THE ATTORNEY

Questions from Readers

By Beth Sufian, Esq.

The following questions have been asked by readers of CF Roundtable. *Questions asked by readers are never disclosed without the agreement of the reader and information never will be published that would allow anyone to identify the reader who asked the question. Nothing in this column is meant to be legal advice about your specific situation. If you have additional questions please contact the CF Legal Information Hotline at: (800) 622-0385 or by e-mail at: CFlegal@cff.org. The Hotline provides free and confidential legal information to people with CF, their families and their CF Care Center teams. The Hotline is proudly sponsored by the CF Foundation.*

1. When can a provider send an outstanding bill to collection?

In the current economy there are many health care providers who have stepped up efforts to collect medical debt from patients. Many states have laws limiting the number of times a bill collector can contact a person about a debt. Some states have laws that limit the tactics a bill collector can use to try to collect a debt. For example, in some states a bill collector cannot go to your home or office to try to collect a debt. Generally, a provider can send a debt to a collection agency at any time after the debt is incurred by the patient. Therefore, people who know they will have trouble paying the co-pay for an office visit or for a hospital stay may want to discuss payment options after the visit occurs. Hiding the bill in a drawer is not a good solution, when a person cannot pay an outstanding balance.

BETH SUFIAN

A person can always ask providers if they will allow the person to pay an outstanding balance on a bill over time. Agreeing to pay an outstanding balance over time is a good way to avoid having a debt sent to a collection agency. Most CF care providers know that people with CF have high medical costs. Many CF providers often are willing to arrange a low monthly payment plan toward the eventual payment of a debt. For example, a person could explain that they are paying $25-a-month toward a hospital bill, a physician bill and a pharmacy bill. If there is an outstanding balance with a radiologist, the person could show that they already are paying $75-a-month toward other bills and would be able to pay only $20-a-month toward their outstanding radiology balance. The radiologist does not have to agree to accept the monthly payment. However, most providers would rather accept a monthly payment than send the debt to collection and, perhaps, never get paid at all or have to pay the collection agency a fee for collecting the debt.

Typically, a person who is making payments to a provider under a payment plan will not have the debt sent to collection. A provider can refuse to accept a payment plan if he feels the person can pay the balance of the bill or for any other reason. There is no federal law and no state law preventing a provider from sending an outstanding balance to a collection agency. However, there are some laws that prevent the collection of some co-pays for people on Medicare and Medicaid. If a person's circumstance changes and the person is not able to make the payment one month, the provider should be notified and told the person cannot pay that month.

In addition, some providers will settle an entire bill for less than is owed, if the provider will be paid a portion of the money up front. For example, if $100 is owed and the person agrees to pay $80 to settle the account, many providers will accept the $80 as payment in full. Collection agencies are often interested in trying to settle debt cases because the agency usually gets paid a percentage of the amount that is collected. It is better for the agency to settle a debt for $80 and get some payment for their services rather than to get nothing because a person cannot pay $100. Typically, a collection agency will discount the bill by 20% if the person pays 80% of the full amount owed. The collection agency wants to be paid their fee and, typically, will get their fee only if they collect the money owed to the provider or most of the money owed to the provider.

If a person does not negotiate or pay the collection agency within a

certain period of time, the collection agency may report the debt to a credit bureau. Many people are concerned about having a debt reported to a credit bureau. They are concerned their credit rating will be affected if an outstanding debt is reported to a credit bureau. Typically, a collection agency will report a person's debt to a credit bureau only if there has been no indication a person will pay their debt. It is helpful to discuss the possibility of paying the debt with the collection agency. It is best to deal with the provider, initially, if there are issues with paying medical bills.

2. Are there new rules regarding traveling on an airplane with a portable oxygen concentrator?

The FAA has issued a final rule regarding travel with portable oxygen concentrators. The final rule is found at Section 382.133 and requires that all carriers conducting passenger service (passenger capacity of 19 or more) must permit someone with a disability to use a ventilator, respirator, continuous positive airway pressure machine, or an FAA approved oxygen concentrator, unless the device fails to meet the FAA requirements for Medical Portable Electronic Devices and does not display a manufacturer's label that indicates that the device meets the requirements. However, until labeling is implemented there are rules to determine whether the device is acceptable. For more information, go to: FAA.gov. Also, it is a good idea to speak to someone at the airline to discuss traveling with a portable oxygen concentrator. In addition, it is important to discuss traveling with oxygen with your treating physician. ▲

Beth is 42 and has CF. She is a Director of USACFA. Her contact information is on page 2. You may send her questions of a legal nature that are CF-related.

Information from the Internet...

Compiled By Laura Tillman

This issue brings a potpourri of articles from the Internet

NEWS RELEASES

Insights Give New Hope Against Cystic Fibrosis

White blood cells called neutrophils play an important role in the severe lung damage suffered by cystic fibrosis patients, according to U.S. researchers. Their findings may open up new targets for treatment of CF. It was long believed that neutrophils attacked and destroyed bacteria that became trapped in the excess mucus that collects in the lungs of CF patients. But something goes wrong, and neutrophils quickly die in the lungs, releasing tissue-destroying enzymes as they do. However, when researchers analyzed fresh neutrophils from CF patients' sputum, they found that signals from the patients' lung tissue were reprogramming live neutrophils with conflicting messages that "are messing them up completely." As a result, the neutrophils may release large amounts of an enzyme called human neutrophil elastase, which destroys the elastic fiber of the lungs. In healthy people, neutrophils never release this enzyme into nearby tissue.

SOURCE: Stanford University, news release, March 16, 2009
http://tinyurl.com/cqk3fe

CF Foundation Finds Collaborator to Ensure Development of Important Pancreatic Enzyme

The CF Foundation announced that it will collaborate with a pharmaceutical company to ensure development of an important porcine-free pancreatic enzyme replacement therapy. In a Foundation investment worth up to $3 million, Alnara Pharmaceuticals Inc. will complete the ongoing Phase III long-term safety study of liprotamase.

Following completion of the study, Alnara will complete clinical and regulatory activities needed to file a New Drug Application with the U.S. Food and Drug Administration. The company is committed to bringing this important product to patients with CF. Liprotamase was previously in development with Altus Pharmaceuticals Inc. and was known as ALTU-135 and Trizytek. Liprotamase, which has been in development since 2002, was originally discovered and tested by Altus Pharmaceuticals. Unfortunately, due to financial difficulties, Altus discontinued development in January 2009. Unwilling to let this important therapy languish, the Foundation retrieved rights to the potential drug from Altus, maintained its clinical progress and searched for a company to see the enzyme through the approval process. The Foundation selected Alnara because of its unique experience with the pancreatic enzyme therapies: two key members of management were involved in developing the original product at Altus. Alnara also plans to develop a pediatric liquid form of the pancreatic enzyme.

Liprotamase is unique and important because, unlike other pancreatic enzymes currently available, it is derived from non-porcine sources. In addition, liprotamase offers potential advantages over the current standard of care. Today, people with CF must take multiple capsules—sometimes four or five—with every meal or snack.

In contrast, with liprotamase,

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SPIRIT MEDICINE

The Spirit of Stuff

By Isabel Stenzel Byrnes

In this edition of *Spirit Medicine*, I'm going to tackle a potential spiritual remedy for a very personal habit of mine. Okay, here's the confession: I've been accused by more than one close friend of being a pack rat. I've tried to live a spiritual life: one that values emotional and spiritual connections more than material things. But somehow, in this consumer culture, I have managed to collect tons of stuff.

Recently, my husband, Andrew, and I found some spare time to organize our home after many years. Like an episode of TLC's "Clean Sweep", we dug deep into the closets, drawers, bookshelves and, God forbid, the garage. We went through clothes, books, collectibles, sports equipment, stuffed animals, dishes, and the list goes on. It occurred to me that we have a tremendous amount of stuff for just two adults. I had to confront my hesitation over the years to get rid of unneeded belongings. I've been great at making excuses: I didn't have the energy, it was too dusty, it was un-environmental, and it was not a priority. But this year I faced a truth: I am attached to my stuff. It is agonizing for me to toss things away. It isn't the material object that I care about. I have a hard time letting go of the meaning and spirit of my stuff.

I wondered why I collect so much crap. Honestly, I think my cystic fibrosis has played a role in my attachment to stuff. When I first went to CF camp, I remember the night when the older girls' cabins did their skit. Instead of a silly performance or lip sync, they played an audio tape for the audience of their cabin mates in prior years, singing and gabbing like typical teenagers. Many of the girls had died, and those remaining started to cry on stage. Soon, most of the audience was wiping tears.

Since then, I've known my life would be shortened. I wanted to treasure everyone I met and everything that meant something to me. So, I also started to record my CF camp friends over the years on tape. I've kept those tapes. I also have created voluminous scrapbooks packed with photos for every year since high school, some weighing ten pounds. If I became sicker, I wanted to look back on the good times. If I died, I'd leave something behind. I could relive the past whenever I wanted to. Thirty-seven years later, my home is packed with memories.

I keep many things because everything has a meaningful story. For example, the stuff in my house documents my story of sickness and wellness. I have curtains and mosaics I made when I was on disability. To get rid of those items would feel like I was denying my creative energy at that phase of my life. Likewise, I have symbols of my healthy transplanted life: medals from the Transplant Games, bibs from races, an old pair of hiking boots. I'm keeping these things as reminders of my donor's gift.

> *"The stuff in my house documents my story of sickness and wellness."*

ISABEL STENZEL BYRNES

Everything I own connects me to someone. I am inspired by the words of Anais Nin, "The value of a personal relationship to things is that it creates intimacy, and intimacy creates understanding, and understanding creates love." I am a grateful recipient of numerous gifts from generous friends. I treasure a blanket a friend knitted for me, a mosaic teapot my friend made for me. These things exude the creative spirit of my friends. I feel

their loving patience as they carefully made these things for me. Some of the strangest things represent my attachment to people. My CF friend, Nahara gave me an old lamp when I moved into my first apartment. It has a faded and dented shade and is totally out of fashion, but I can't get rid of it. It gives me a sense of security and contentment that she's still with me.

Some stuff I am eager to get rid of. Immediately after my transplant, I wanted to purge all reminders of CF from my house. I packed most of my medical supplies and shipped them to an international charity. Yet, when I was recently cleaning the drawers in my office, I came across an oxygen tubing connector and a "red cap" that I used for home IVs. I held these objects up and examined them - reminders of a life I never wanted but got, a battle I've fought and won so far, of a life I might have again someday. I threw them away.

I freely purged some things from my ancient past that held little meaning. For example, I got rid of the blazers with shoulder pads. Time to throw out my size 4 prom dress! Also, Andrew and I recycled dozens of cassette tapes. Goodbye Culture Club, Tiffany and Duran Duran. These historical items reminded me that I'm growing older. What a privilege.

Some stuff causes regret. When I opened some cabinets in the garage, I uncovered the boxes I had saved from graduate school that just took up space and was fodder for a house fire. I went through these boxes of notes, readers and textbooks, remembering how I once knew so much about epidemiology, biostatistics, psychotherapy and social work. I was overcome by great sadness. These pages spoke to me, reminding me of what could have been if I had not gotten sicker - the career I could have had, the income I could have earned, the professional I could have become. I held onto this stuff for decades, hoping and wishing I would use this knowledge someday in a job. With my husband's reassurance, everything went into the recycling bin.

My greatest material attachment comes when I find things my deceased friends have given me. I've kept one earring of a pair given to me by a CF friend who died years ago. Her father gave me a little plastic owl toy that moves and squawks. It has no purpose except silliness and a deeper reminder of his love for her. I kept a rusty Christmas ornament from a CF friend who died two months after giving it to me. I am holding on to these things because I'm holding onto their love. But holding on hurts.

After my close call with death, before my transplant, I realized my husband and family would be burdened by all the stuff I would have left behind. Everything that belonged to me might cause pain for them as a reminder of my loss. I don't want to do this to them, so I have tried to buy less and rid my house of extra weight. I regret materialistic binges I had before transplant. I mean, did I really need that china or Disney kitsch? Those purchases offered a brief glitch of happiness. Now I'm lucky to find other sources of joy.

I am finding that less stuff is good for my spirit. Eliminating clutter in my living space has created a sense of tranquility and coherence. I display only those items that are representations of a higher or deeper reality such as nature photography or art. By limiting sensory input to a comfortable level, and only surrounding myself with beautiful things I love, I appreciate more what I own. As I donate some of my stuff to the Goodwill, I am further comforted by believing that I am passing on the spirit of my stuff to other people who need it more than I do.

As I downsize now, I am working on extricating the spirit from my stuff. The memories I have - the stories, the connections, the past, the love, the regrets - live inside of me, not in the metal or plastic form of the object filling my home. I keep my memories alive by sharing them verbally or through writing. I need to accept that I don't need my stuff to hold onto memories. I remember that the moment of those memories is what really matters, and I can't live in the past. In fact, my 93-year-old grandmother can't remember much these days. She is teaching me that what I think is so important to remember is actually not. In the end, if I forget, it's okay.

Detaching memories from stuff also means accepting the losses I have experienced. I must let go of my need to keep my friends alive through the stuff they gave me. That is too great a burden. I find comfort in what psychiatrist Irvin Yalom wrote: "People die when I die, when everyone they've touched dies. They don't disappear into nothingness, they enter a different realm which we don't understand, they re-enter this world's energy, or become part of everything, or these bits of memories reattach and complete themselves in an even better place than here." There is something so much greater than our own grasping to objects that will connect us to our loved ones. I tell myself that no object - no material form - represents what I have inside from people in my past. My love for the people who gave me things is no greater or no less if I hold on to or give up these things. Likewise, my passionate grasping onto my life - past, present and future, is unchanged by the goods I possess. The only truly meaningful stuff I can own is stuff that enhances a deeper spiritual appreciation of life. It doesn't take a lot. ▲

Isabel Stenzel Byrnes is 37 and has CF. She lives in Redwood City, CA. She invites you to share your spirit medicine.

SPEEDING PAST 50...

I Am An Aunt

By Kathy Russell

The decision to become a parent is a very private decision that frequently is discussed by people who have no part in it. Many young people find that there is pressure from family and friends about when they should have children, how many children they should have, or even if they should have children. Most of us who are older have been through this.

Some of us don't want children and never did. We may be the rare beings, but I know there are many of us who have CF and who don't have or want children. We marry, have a long and happy life and never regret not becoming parents. I know many couples who have been married many years, without children, and they are quite content with that. I also know many couples who have children, either the old-fashioned way or by adoption. They are happy with their decisions, too. The freedom to choose is wonderful.

I am not a mother, but I am an aunt. I have been an aunt since I was 27 months old, when my eldest sister had her first baby. My brother's wife had their first child four months after that and the babies just kept coming. Between the two couples, they had seven babies in my first 10 years.

It seems as if there always were kids around our house. My mother was very good about watching her grandchildren anytime that the parents needed a break. Once I was about 13 or 14, I did a lot of baby-sitting for our neighbors. As a result, I spent much of my younger years looking after other people's children.

By the time I was an adult, I was tired of kids. I thought they were nice, but I wouldn't want to have one in my home. It was rather the same way I felt about wild animals – they were okay to look at but I wouldn't want one at home.

When I got married, I was fortunate enough to marry a man who didn't have a burning desire to be a father. We found that we were quite happy being just the two of us. We enjoyed each other's company and liked the freedom we had without kids. We each like our privacy and time alone. It would be hard to find that kind of time if we had kids.

We still had many nieces and nephews around. Also, we had six godchildren who were an important part of our life. We could enjoy them when we wanted to and we could send them home when we were tired of them. We didn't have to endure childhood illnesses or teacher conferences. We could enjoy school programs and celebrations of the happy times in their lives. It was the best of both worlds.

KATHY, 3, AND HER SISTER, IRENE, AGE 8, WITH A BABY NEPHEW. (CIRCA 1947)

There were several times that we had a child live with us. Those times were of short duration and were bearable. I did find that I didn't get enough rest when we had children in our home, however. It always seemed as if there were too few hours to get everything done, because the kids' needs had to come first. Paul was still working and most of the work of taking care of the kids fell to me. I was happy when it was time for each of them to move on.

During the years that I worked, I worked in several different areas of hospitals. I spent the most time in pediatrics and in Newborn Nursery. I enjoyed pediatrics, where I was working with school-age kids, but I got burned out because most of the kids were "high risk". Nursery was much more my cup-of-tea. I loved taking care of a baby for the first three to five days of life. Rarely did a baby stay with us longer than five days. It was long enough to get one's fill of cuddling with a sweet baby, but not long enough to get too attached to any baby. The babies were small enough that they were easy to lift and to care for. Over the years, I took care of thousands of babies and I really enjoyed it. If I had any bit of maternal instinct, it was satisfied with all those babies.

The nursery was a very clean place to work. It was air-conditioned, at a time when most parts of the hospital were not, and no one who was ill was allowed to stay on the area. Even at that, I cultured Pseudomonas aeruginosa (Pa) for the first time in my life while working there. There were not the antibiotics to deal with it then and people were as afraid of it as they now are of B. cepacia. Being colonized with Pa was considered to

be a death sentence. Since more than 35 years have passed and I still have it, I guess it was more of a life sentence than a death sentence.

It was while I was working in the nursery that I had a hysterectomy. My husband, Paul, and I had talked often and at length about whether to have children. There was no way to know if Paul was a carrier of the CF gene. This was years before the gene had been identified. Pregnancy was considered to be dangerous for women who had CF. Another concern was that Paul's blood type is "positive" and mine is "negative", which could cause some problems. I was doing fairly well and we didn't want to "rock the boat", so to speak.

After much discussion with my doctor about the various possibilities, we decided that a hysterectomy was better than a tubal ligation. I had been taking oral contraceptives for about eight years and that was not good with my high blood pressure. We did not want Paul to have a vasectomy because there was no reason for him to avoid fatherhood if something should happen to me and he remarried. As long as I would have to have an abdominal incision (laparoscopic surgery was just being tried and was not an option), why not do the hysterectomy and avoid possible future problems? We have discussed this decision, many times over the years, and always have decided that it was a good and solid decision, and it definitely was the right one for me. I never have regretted having it done.

So how did we fill our lives? Paul and I find that most often we are just plain Uncle Paul and Aunt Kathy. Oh, boy, are we an aunt and an uncle! We have 18 nieces and nephews, who have 27 children, and they have even more children. It seems that everywhere we turn there is a niece, nephew, great-niece or great-nephew, etc. That is fine with us. Again, as with our godchildren, we can enjoy them and send them home.

Recently one nephew, who lived with us while he was in high school, was here to help Paul with several projects around our place. I know that both of them enjoyed their time together. Another nephew, who spent his days with us while his mom worked when he was a preschooler, came to help cut up trees that fell during the winter. His wife wanted to learn about canning at home and I was able to give her a lesson on the basics, while their baby slept. After the fellows were done and the baby woke up, we all had a great dinner together. It is so nice to have the kids come back all grown up. It is a pleasure to be able to interact with them as adults.

Each person has to find the answers to the questions of parenthood that they can live with. What worked for me, may not work for you. Many people who have CF have children the "old fashioned" way. Others may adopt either an infant or an older child. Now there are not as many roadblocks in the way, to keep people with CF from adopting, as there were when I was young. Some people adopt children from other countries. I know of a few who have done that and they have been very happy with their children.

If you decide to have a child or to adopt a child, I hope that you enjoy your child or children. Also, I hope that you and your family are able to enjoy many, many happy years together. If you decide to forego having children, I hope that you will have a life that is as full and happy as mine has been with my husband for more than 44 years.

All that Aunt Kathy wishes for you is a healthy, happy life…no matter what your choices are. ▲

Kathy is 65 and has CF. She is a Director of USACFA. Her contact information is on page 2.

INTERNET *continued from page 5*

patients can take one small capsule with every meal or snack.
http://tinyurl.com/cdu5l5

FDA Approves Pancreatic Enzyme Replacement Product for Marketing in United States

Creon designed to help those with cystic fibrosis, others with exocrine pancreatic insufficiency

The U.S. Food and Drug Administration announced that it has approved Creon (pancrelipase), a pancreatic enzyme replacement product designed to help patients with cystic fibrosis digest and absorb nutrients from foods. Creon is the first FDA-approved delayed-release pancreatic enzyme replacement product to be marketed in the United States as a result of the agency's unapproved drugs initiative. Creon, which contains a mixture of digestive enzymes extracted from the pancreas of pigs, helps patients lacking the enzymes needed to digest fats, proteins and sugars from food. Creon is approved for use in pediatric and adult patients. The FDA had required the manufacturer of Creon to submit, and the agency has approved, a Risk Evaluation and Mitigation Strategy (REMS), which includes a Medication Guide, to advise patients on risks associated with high doses of Creon, and the theroretical risk of transmission of viral disease from pigs to patients. A rare bowel disorder, called fibrosing colonopathy, can result from a patient's high-dose use of Creon. While this condition is serious and may require surgery, a patients' chances of having this condition may be reduced through their adherence to dosing instructions in the labeling. Creon may be sprinkled on food. Because Creon is a delayed-release drug, patients should never crush or chew the capsule as this would cause an early release of the enzymes and a reduction in enzyme

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WELLNESS

Parenting With CF: A New Normal?

By Julie Desch, MD

As I sit to write the *Wellness* column today, it is the last week of school and I am to write about "Parenting with CF." How good is life? Finally, something I know a little bit about!

Do you remember how you felt on the last day of school when you were a kid? Well, as a parent, you get to experience it all over again. Today, I am 24 hours away from weeks on end of no homework, no packing lunches, no craziness trying to get out of the house on time in the mornings, and no schedules!

But I digress. My purpose is to talk about maintaining wellness while being a parent, so I guess I should stick to that. First, you should know — it isn't easy. In fact, it is most definitely the hardest thing I have ever done…and keep on doing.

Don't get me wrong…deciding to become a parent was the best BIG decision I ever made; period. It beat moving from Nebraska to California (that was a big one). It beat going to medical school. It beat retiring from medicine. It beat acquiring more dogs than is legally allowed in my city…don't tell. It was the right thing for me to do. And when I decided to do it, I had NO idea what I was getting into. It is only in retrospect that I know how right it was.

But you are going to know, because I'm going to tell you. As I would do when coaching anyone who faces a big decision, if you are contemplating becoming a parent and you have CF, you really need to take a PRO/CON approach to the decision. I'll try to take a balanced approach to this from my perspective, but it could (will) be different for you; so consider my words with a large grain of salt. As I list my pros and cons, I'm going to try to connect them to the "wellness" ramifications that they hold.

First, there are the emotional benefits:

Love

This is hard to convey in words. The love you feel for a child that is yours is truly indescribable. I had no clue that I could actually feel the way that I do about my two sons. For me, it feels like a mixture of unconditional adoration, fierce pride, ferocious protection, intense joy, equally intense fear (of loss), and immense responsibility. It isn't anything like the love you might feel for a partner/spouse or family member…it's a whole different ball-game.

So how does this feeling benefit your health? Did you read the "connection" article from the last edition of *CF Roundtable*? The study I mentioned in that article showed that people who had the closest connection with other humans were the happiest. And in my book, to be "well" is much more about overall happiness than health (otherwise how could a delta F508 homozygote possibly write about wellness?). Can you be happy and childless? Absolutely. Will it benefit you emotionally to feel the love that I am trying (and failing) to describe? I don't know…but I know it has benefited me.

IN THE HAMMOCK, JULIE DESCH'S CHILDREN, GRIFFIN AND PHOENIX.

Laughter

A sense of humor is a strength. We all know this intuitively. We also know through scientific research that laughter is good for health. Humor and laughter strengthen your immune system, boost your energy, diminish pain, and protect you from the damaging effects of stress.

Is there anything more contagious than the laughter of a baby? I dare you to search for the "Funny Baby Laughing" video on YouTube right now and not laugh… out loud… several times. Your own baby will do this to you, too. Mark my words. Then it just gets better as they get older and wiser. If you don't believe me, live with Curly and Mo (aka Phoenix and Griffin) for a day.

Legacy

This one is a bit more nebulous, for me anyway. But it is definitely an emotional plus. I'm not talking here about a monetary legacy or a best sell-

ing memoir. Instead, this is a feeling that I have as a parent that was missing when I didn't have children; where I want to do the right thing (i.e. take care of myself, not complain, be courageous, etc.) because I want my children to remember me that way. Does this make sense? The act of wanting to model positive behavior for my kids is actually good for me! A win-win, I would say.

Motivation

This is a big upside to being a parent. Take one adult living with a life-threatening illness that demands a rigorous self-care regimen, add one tiny, completely dependent infant, shake gently, and you get…instant motivation! It is not about just YOU anymore. Now, you do what you do not just for yourself (or spouse, or parents, or doctor), but also for that tiny little rug-rat that needs you to stay healthy.

When my sons were very young (preschool age), I remember starting up a running program (again) after being sick. The first few runs of a running program, at least the way I did it back then, were not fun, to put it mildly. I needed to come up with something compelling to get me out there. It was natural then…I kept telling myself, "This is so that I'll be there for high school graduation." Then, as they (and I) got older, it became, "I need to do this so that I can see them graduate from college." Now, it is, "This is to be there for that first grandchild."

The purpose for staying healthy is now much larger than myself. This makes it more pleasant and easier to keep going.

Then, there are a few emotional challenges (and when I say "challenges", I mean "cons"):

Stress

This is a very benign sounding word. Really, what I should say is "overwhelm". It is truly amazing to me how one *tiny little baby* can lead to such extreme fatigue. Now, granted, I started a little late in life. I was 36 years old when my first was born, and 38 when my youngest appeared. It didn't help that when they were 3 years old and 18 months old, respectively, I found myself a single parent rather unexpectedly. Now, this was NOT the plan, and this brings up a great point: You can't anticipate everything that will happen. You may think you are prepared for anything, and then…stuff happens.

> *If you are contemplating becoming a parent and you have CF, you really need to take a PRO/CON approach to the decision.*

It just so happened that this "winter that my life fell apart" was also the worst winter of my life from a health perspective. Imagine that. I needed IVs twice that winter (after only once before in my entire life). The oldest was in preschool and brought home every imaginable virus. I was sick constantly. I don't remember sleeping much.

Not to be the bearer of bad news, but living with a baby, even if there are two of you, is difficult and, at times, very stressful. I highly recommend developing an active stress reducing activity prior to that first diaper change. I also recommend that you and your partner be realistic about who is best able to cope with lack of sleep, what you plan to do when the baby gets sick (hint…run away!), and how to best maintain your sanity…and your relationship.

Guilt

This dovetails nicely with the above. Guilt happens when your child is sick and you can't snuggle or when *you* are sick, and can't be the perfect PTA-mom or dad. It is *really* easy to feel bad when you have to say, "I can't (fill in the blank) honey, I have to do my treatment," as they look at you with those big eyes. Well, maybe you won't; but I sure did, and still do.

Guilt is not good for you. It serves no purpose other than to make you feel bad for something you have no control over. Guilt robs you of happiness and, as I said above, this robs you of wellness. Good news, though. Guilt is optional.

Moving on to the physical benefits:

Snuggling

The best…warm, fuzzy feelings are happening to me just thinking about this one. How healthy is physical contact? We don't know why yet, but I'm sure you've heard what happens to babies when they are not held. Well, the benefits go both ways. Social isolation is a major risk factor for mortality from widely varying causes. Physical contact with your child is as important for you as it is for him or her.

I hear you. Yes, snuggling with a partner is also good for you. The way I figure it, a child adds to the pool of possible snugglers, which cannot be a bad thing.

Tickling

(Read "laughter" above)

Moving

I can't write an article on wellness without a mention of physical activity, now can I? A child, like a dog, forces you to move. In fact, a child forces you to move a LOT.

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FOCUS TOPIC

BECOMING A PARENT WITH CF

Wonderful Surprises

By Lynn Williamson

I am 41 years old. I have a wonderful husband, Darren, and the two most adorable children. Our son, Cameron, is about to turn nine years old. Our daughter, Erynn, just turned one year old in May. Yes, I had her when I was forty. She was a surprise. Cameron was too.

My previous husband and I tried for five-and-a-half years before I became pregnant with Cameron. We both were tested. I did three or four rounds of fertility drugs. We figured it would never happen. So we decided to look into adoption. The day I got the adoption application was the day that I found out I was pregnant. It was the best surprise!

My pregnancy was remarkably unremarkable. I felt great throughout the pregnancy. I loved being pregnant! I did not have any breathing problems or anything else. In fact, I was healthier during my pregnancy than I had been prior to it. I never even had one day of morning sickness! I attributed this to the fact that because I had had so many sinus surgeries, I had not been able to smell anything for about three years.

The only day in the whole pregnancy that I really felt bad and wondered how I was ever going to get up and drag myself to work was the day I went into labor. Cameron was three weeks early. He was big and breach so I had known for about two months that I would need a C-section. I recovered quickly.

I went back to teaching after only six weeks off. The hard part was leaving my beautiful baby boy when I did. I think I cried that entire week at school.

I had always had bad sinuses. That was about the extent of my problems with CF. I had to take enzymes and I had sinus surgeries. At this point, I did not do breathing treatments or, really, anything else. I had already had six sinus surgeries.

When Cameron was five months old, I had to have my gall bladder removed. I was in the hospital for six days. The hardest thing was seeing my baby only once during that time. My parents kept him. Again, I recovered quickly and was back at work only three days after being released from the hospital.

I began to have sinus problems again. I had surgery when Cameron was 27 months old and again when he was 2½ years. I had a staph infection in my sinuses that we could not get rid of. During this time my husband decided to leave.

One month after the last surgery, I had my first ever IV antibiotics. It scared me to death. I stayed in the hospital two days, and then was sent home with home IVs. I didn't have a PICC line. Every two days the medicine blew out my vein and a nurse came out and started another site. My mother stayed with me during this time. She would get up at 4:00 a.m. to start my drip so I could sleep a little more and my meds would be finished in time for me to go to work. Cameron, at 2½ years old, learned how to stop the machine from beeping and even helped get air bubbles out of the line.

One month later, I had the BIG surgery. I have a scar that goes from ear-to-ear over the top of my head. The doctor went through the front of my skull. He scraped out my frontal sinuses and filled them in with fat from my abdomen to form scar tissue. This was supposed to prevent any more bac-

ERYNN, DARREN, LYNN,AND CAMERON.

teria from being able to get into my sinuses. It worked for about six years. I told the doctor that was my one chance at a face lift and a tummy tuck, but he left the fat and the wrinkles.

In 2003, I met my husband, Darren. We were married in 2004. We tried to have a baby. It wasn't working, but we weren't willing to go the fertility drug route. After all, I was 37 when we got married. Darren was happy that we had Cameron. From the day we were married, Cameron chose on his own to call Darren, "Daddy".

As I approached 40 we discussed getting my tubes tied. My health was still good - no breathing problems - and only one sinus infection in three years. It was getting to that time to have my yearly check-up. I mentioned to my doctor that we had been trying for three years to have a baby. She did a test to see if I was ovulating. The test came back that I was not. Oh, well. I guess that decided that for us, right? Wrong!

The next month I decided to take a pregnancy test. I don't know why, I just did. WHOA!!! There were two pink lines. I took two more. Yep! I was pregnant at 40!!! Now what?

I was anticipating the same easy, fun pregnancy as I had had with Cameron. WRONG again!!! I had morning sickness, afternoon sickness, evening sickness, and every time in-between. Obviously it is not smell that makes you sick during pregnancy. Remember, I haven't had a sense of smell for about 10 years now, but I sure was sick. Then the breathing problems started. At about two months pregnant, I went into the hospital for the first time. I had bronchitis for the first time. I had a PICC line for the first time. I was scared.

I am allergic to so many antibiotics. The ones the infection was sensitive to, I was allergic to. The ones I could take while pregnant, the infection wasn't sensitive to. I was put on Vancomycin. I went to school with my red and purple PICC line tentacles hanging out of my sleeve. After I finished the round of antibiotics, the PICC line came out. Unfortunately, the next month I was back in the hospital for another round of bronchitis and another PICC line.

When I went in again one month later, it was decided to keep the PICC line in until I delivered. I was only half-way through. At 20 weeks, I had to have surgery for a preexisting condition. I was given a spinal and had surgery while I was awake. That was an experience!! I went in the hospital two more times before it was over. I was forced to take leave from work five weeks early. Toward the end, I was able to sleep only in our recliner, because I couldn't breathe lying down.

I went into labor 3½ weeks early, or so I thought. I knew I was in labor. We all loaded up and headed to the hospital - 45 minutes away. The only problem is that we live on the north shore of Lake Ponchatrain and the hospital is across a 25 mile causeway. Wouldn't you know it; they had the draw bridge up inspecting the bridge. Luckily, my husband had the number to the bridge operator just in case this happened, bless him. They lowered the bridge and we rushed to the hospital. It was false labor, and they sent me home.

Two days later, I knew my lungs were bad again. I went back to the hospital and they admitted me. When they put me on the monitor, I was having contractions! Well, now they had to stop my contractions for two days to get my lungs ready for the C-section.

Another two days later, we met our daughter, Erynn. She had put us through a lot to get her here. It was absolutely worth every single thing I went through!!! She is so beautiful. She has her daddy wrapped around her finger. Her big brother thinks she hung the moon. Whenever he walks in the door, she just lights up and runs to him. She has her grandparents in the palm of her hand. But my little angel came with a price.

I had to have another BIG sinus surgery when Erynn was only six weeks old. The doctor had to undo what had been done before and more. He took out part of my septum and the middle turbinates on both sides, and cut new drainage holes lower in my sinuses. I was basically unconscious for about a week because of the pain. I missed the first two weeks of school.

It's coming up on the one year anniversary of my surgery and I still have to go to the doctor at least once a month to have my sinuses cleaned out beyond what I can do at home. I never had bronchitis until I was pregnant. I never coughed. Now I cough all the time. I do Xopenex, Pulmicort, Pulmozyme, and am adding hypertonic saline. I have crackles in my chest and thickening of the bronchial tubes in my lower lobes. I don't know what will come of all this, if any of the damage can be reversed, or if now it will all be about damage control.

My husband and I have decided to start a cardio exercise program to help build my lung function and stamina. Hopefully, this will help prevent anymore damage and maybe reverse some of what is there. I believe with all my heart that God gave me the opportunity to be the mother to the two most wonderful souls on earth and wife to my best friend. So I have to believe that He will allow me to see our children grow up and have families of their own, and to grow old with my husband. I do know that it will look different from what I had imagined a year ago. But, isn't that the best surprise of all? ▲

Lynn is 41, has CF and is a teacher. She and her family live in Covington, LA.

No Parenthood Planned

By Susie Baldwin, MD

From the time I was a kid, even before I knew I had CF, I thought that one day I would want to adopt a child. I couldn't understand why people made more babies when there were already so many living children who needed homes.

As I grew older, I learned to appreciate and understand why people choose to bear their own genetic children. But bearing a child of my own wasn't high on my list of things to do. My focus remained firmly on my education and my career.

SUSIE BALDWIN PLAYING AT THE BEACH.

From the time I became sexually active at age 19 (I was a late bloomer—despite all the political controversy about teen sex, the average age of first intercourse in the U.S. is 15), I was extremely conscientious about using birth control. In the dawn of the AIDS era, that meant consistent use of condoms as well as going on the pill.

I finished medical school and began a residency in obstetrics and gynecology, within months experiencing the pleasures and occasional horrors of delivering hundreds of other people's babies. (On my very first night being a doctor, July 1st of internship year, I was monitoring a patient in labor. Her uterus ruptured, and her baby wound up in her abdomen. Her placenta fell to the floor. It was a devastating introduction to my career, though the mother and the baby were saved by the speed, skill, and grace of Dr. James Machiulla). Most importantly for me, I learned to help women who weren't ready or able to be mothers to safely prevent or terminate their pregnancies. I became a family planning expert, an abortion provider, and a preventive medicine specialist. Though I got married at 29, having kids of my own remained a vague plan for the future.

After I turned 31, I started having increasing difficulties with my health, and dealing with CF became more of a daily battle. I realized during a bout of illness at age 32—the first time a winter virus made it difficult for me even to walk from the bedroom to the bathroom—that it was challenge enough for my husband and me to take care of me, and maintain our lives and careers. It would be incredibly difficult, if not impossible, to also properly care for a child, much less incubate one for nine months. While my doctors told me that I could safely get pregnant, and referred my husband for genetic testing to see if he was a CF carrier, the right time never came to explore these options more fully.

For many women and couples, procreation is a fundamental part of building a family and a life together. People for whom pregnancy does not occur naturally often go through tremendous effort, expense, and heartache to find their fertility. For many women and in many cultures, motherhood defines womanhood. I did not hold these beliefs nor, fortunately for me, did my husband—though we did not explicitly discuss any of these matters, that I recall, before we pledged to live our lives together.

Not becoming a mother, for a heterosexual woman in a relationship, typically requires paying a lot of attention to birth control, for a long time. The average woman who wants to bear two children must practice contraception for three decades; without contraception, women experience on average 12 pregnancies and bear eight children. Birth control pills, emergency contraceptive pills, the patch, the ring, condoms, shots, cervical caps, intrauterine device—I have used them all, with great success. I've been so successful at preventing my own pregnancy that I sometimes

wonder, if I had ever tried to get pregnant, would I have been able to. At this point, I'll never know.

Regardless, I like to think that my personal experience with nearly every form of birth control on the market enables me to provide better contraceptive care to my patients. And I like to think that my active decision to not become a parent has been an appropriate and responsible one.

As my health declines with the years, I am able to focus my energies on staying or getting well and contributing to the world through my career and my activism—fighting for others' reproductive freedom and health care for all. I am able to enjoy my nieces and nephew and countless friends' kids, but I can sleep in on the weekends, getting the rest critical to living with CF, and not worry that anyone is waiting for me to make them breakfast or take them to soccer practice. I can politely decline invitations when a child has the sniffles or when I'm not feeling quite up to a social event. I feel good about not contributing to the overpopulation of the planet and the over-consumption of scarce resources.

This is not to say that I don't respect or admire those with CF who have had the good fortune to bear or adopt children—and I do sometimes wish I could contribute to the planet another being with my husband's kindness, wit, and beautiful long eyelashes.

Still, I am content, in my own life, with being a mother of sorts to my two dogs and three cats, all adopted from shelters and the pound. And still I wonder…why do people breed dogs and cats when there are so many animals living that need homes? ▲

Susie is a physician who is 42 and has CF. She lives in the Los Angeles hills with her husband and animals. You may contact her at: susie.baldwin@gmail.com

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.

A Parent's View 29 Years Later

By Anne Williman

On August 11, 1980, my life changed forever.

"Anne, we have a little girl who wants to come home with you," the social worker told me. "Can you come meet her tomorrow?" I could hardly stammer out an answer. My husband, Jon, and I had been trying to adopt a baby for three years, and now it was finally happening.

We were introduced to our new daughter, whom we named Amy, the next day. She was four months old and perfect. The day after that, we brought her home. And life was beautiful.

That was 29 years ago. Plenty has happened since then, including the adoption of two more babies, both boys. Joe arrived four years after Amy, and he was three weeks old. We had to wait another eight years after that to get Jesse, whom we got straight out of the hospital at the age of five days.

So with the perspective of 29 years of parenting three kids and dealing with my CF, I can say it has been the most difficult thing I've ever attempted. Parenting has given me the deepest pain and the greatest joys of my life. And I've never felt useless or bored!

I quickly found out that being up at night a lot with crying babies did not agree with me at all. Fortunately, my children never did much of that, and I was able to get a good night's sleep most of the time.

I worked to keep my health care going in spite of the radical changes in our lives. I remember Joe swinging in his baby swing while Jon did my postural drainage (PD). (This was before the days of *The Vest*®.)

One day when Amy was about three, she sat down next to me on the couch and began gently patting my calf. It took me a few minutes to figure out that she was just duplicating what she saw her dad doing to my chest every night with the PD.

About that time, I was admitted to the hospital for my first clean-out since she had arrived. My doctor and the hospital were two hours away from where we lived, so I didn't see her for a week. Then after Jon brought her to visit me over the next weekend, we were separated for another week. It was agony for me to be away from her, after spending every night with her ever since she'd arrived.

Finally the day came for my discharge. Even though I was 29, I had been a patient in the children's hospital. (This was before adults were transitioned to adult CF clinics and hospitals.) As Jon and I walked out, hands linked with Amy's, the receptionist smiled sweetly at our little family.

"You must be glad to be taking your little girl home," she gushed. Jon and I exchanged glances, then just grinned and nodded at the woman.

I was thrilled to be home, but Amy acted very strangely. She didn't want to be near me. For the next few days, she continued to be distant. Then one day when we were sitting on the couch, she told me, "Grace's mom is in the hospital."

I was startled. Grace was our dog. What was Amy talking about? Then it began to sink in. She meant me! I took her tiny hand in mine. "Then we'll have to take very good care of her until her mom is home." We talked about how lonely and scared Grace must feel.

From then on, Amy began working her way back to normal. I promised myself then that I would not go back into the hospital again until my kids were older and could handle the separation better.

My health remained pretty consistent, and I was able to keep that promise. Of course, my kids grew up seeing

FROM LEFT: SON, JESSE; SON, JOE; SON-IN-LAW, CRAIG; DAUGHTER, AMY; HUSBAND, JON; AND ANNE WILLIMAN.

me do nebs, down pills, drive to doctors'appointments, and discuss PFT results. The hospital stays came later.

In some ways, they had to grow up too quickly. They all fixed their own breakfasts and lunches at an early age. When I was sick, they even had to prepare my meals! They helped with housework, with laundry, and with my home IVs, and there were times when they asked Jon if I were dying. During those periods, when I was the sickest, their behavior tended to get worse, which stressed me out more.

Then there were the more subtle things. I was one mom who didn't volunteer at the school, serve as a field trip chaperone, or attend PTA meetings. Just trying to keep up with my family and my health were all that I could manage.

Sometimes even that was a stretch. Too often, there were times when I couldn't even manage cooking dinner or running the sweeper. But Jon never complained. If I couldn't do it, he was always willing to get a meal on the table, wash dishes, or do whatever housework I couldn't. It takes a special person to be married to someone with CF, especially if you add kids to the mix.

Before adopting, we had discussed the possibility that he would be raising kids alone. He assured me that he was willing to do so. We agreed that for us, raising kids was something we definitely wanted to do.

Jon was the one who usually put the kids to bed at night. He often took the kids to their activities, and he served as the Cub Scout leader. He always had to do more than your average dad.

When the kids got old enough, they got into sports. As often as I could, I attended their games, which at various times included soccer, football, basketball, baseball, rugby, and roller blade hockey. Then there were track, gymnastic, and swim meets, Tae Kwan Do contests, horse and dog shows. Not to mention Amy's dance recitals. (She took 12 years of ballet.)

I did learn that sitting out in the rain, even to watch Jesse play in his football game, was not wise, since I ended up getting sick enough to land me in the hospital. After his freshman year of high school football, I stayed home when the weather was too bad.

I also survived many years of Cub and Boy Scout meetings and award ceremonies. (The camp-outs and hikes I left to Jon.) There was also 4-H, which meant more meetings, helping kids complete projects in things like cooking, dogs, writing, photography, childcare, and nature, and taking them to judging at the county and state level.

I managed to live with an assortment of pets including a horse, hamsters, fish, guinea pig, snake, rabbits, and six dogs (not at the same time!).

Of course, there were all the holidays and birthdays to celebrate. We treated the kids' adoption days as holidays, too, giving them gifts and doing something together as a family. I have to admit I didn't enjoy the sleep-overs when the kids stayed up all night.

We always took a family vacation. Over the years from our home in Ohio, we visited Maine, southern California, Florida, Texas, Missouri, Wyoming, Canada and most places in between. The kids did their share of complaining about the trips, especially the museums I insisted we check out. (They still roll their eyes at the mention of the Shoe Museum in Toronto.) But now, the older two talk of traveling when they have families.

Of course, for me vacations still meant nebs, The Vest, pills and the rest, but I managed.

Naturally, parenting also brings times that are anything but fun. Take emergency room visits, and please take them far away, because I've had more of my share of them between the three kids. The first was when Amy was about two. We were Christmas shopping in a mall, where Jon was looking at handsaws for his dad. Amy somehow stood up in her umbrella stroller and tipped it over backwards, gashing open her chin.

As her shrieks filled the store and blood gushed down her coat, Jon decided he better put down the saw he was holding! We rushed her to ER. I couldn't bear to go in with her as she was stitched up, but Jon stayed with her. I had no idea that day just how many times I would be taking kids for the same procedure. Now, a round of stitches is nothing.

For a while, our kids seemed to be competing about who could get the most stitches. One kid was sledding when he crashed into a metal well cap, slashing open his forehead. Another had a "friend who didn't know how to play golf" hit him in the head with a golf club as he swung at the ball, cutting away so much flesh that you could see the child's white skull. Someone washing the car fell and slit open a knee on the license plate. Over the years, I think each child received more than twenty stitches. (And I know one of them got way more than that, since he received over that number just one time when he cut open his arm all the way down to the bone!)

Of course, with all the sports our kids have participated in, there have been plenty of injuries. Why does it always seem to be my kid lying out there on the field after the play? We've dealt with a broken nose, serious concussion (took the child out of play for most of the season), sprained knees, ankles, wrists, shoulders, fingers, necks, and probably a few parts I've forgotten. Fortunately, nothing too serious.

I have to mention the many times they were sick, where I stayed up all night with a feverish or vomiting child. I think I spent almost as much

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Stepping Into Motherhood

By Debra Radler

Motherhood. I guess you should approach it with the same amount of courage you would use to jump out of an airplane, because it really is an exercise in blind faith. The greatest difference, of course, is that an airplane jump is over in a few minutes. Motherhood never ends.

NICOLE, DAVID, ADRIAN AND DEBRA.

I decided a long time ago that I would never actively pursue motherhood. If it happened by fate, then I would accept all that came with it. But if left to choice, I would opt out for reasons that are rather obvious to anyone living with cystic fibrosis.

Most of my friends are mothers. My brother and sister-in-law have triplets. I fostered a special needs boy for almost a year when I was in my 20s, was what I would like to call a "pseudo-stepmother" for 10 years to three teenagers while in a previous committed relationship, and I am now a full-fledged stepmother in my new marriage. I know the commitment that is involved in full-time mothering. And for as much as my heart may have yearned a time or two for an adorable little something to love more than life, my mind always recognized the reality at hand.

I knew that I was not up to the task. I knew my body, I knew my personality, and I knew my limitations. The combination of the three left me confident that motherhood was not for me.

First and foremost, there is the carrying and the bearing of the child. I could not imagine an added stress to my system. I couldn't see myself performing chest PT and postural drainage, at eight or nine months pregnant, while gravity is forcing a baby into my lungs. I don't think I would want to deal with all the emotions involved with worrying about the baby's health, and my health, and whether I could stay healthy enough for the delivery, let alone through the delivery. And, of course, there is the crème de la crème worry…would I live long enough to raise my own child into adulthood? It was all too overwhelming for me.

Secondly, there are the first three years or so of the baby's life, when he is completely dependent on me. We can call them the blurry years, filled with sleep deprivation, diaper duty, cuddling the one with colic, wiping noses and kissing boo boos. They are joyful and tiring and stressful and exponentially fulfilling, but they are years that may have been stolen from me, had I chosen to indulge. I don't know that my health would have allowed me to give tirelessly with little time for my own safekeeping. I wonder how many infections I would have caught from my child as he built his own immune system, and I struggled to keep mine strong. When I think of these years, I become more certain that the carrying and bearing would have been the easy part.

Thirdly, there are the adolescent years filled with soccer games, school recitals, homework, carpooling, packed lunches, scheduling and running, sleepovers and driving, driving, driving. The parental demands are endless. I try to see myself getting up early enough to fit in all my treatments, get everyone to their required destinations, keep up a house, take care of the dogs, manage a career, juggle doctors' appointments and activities, in time to fit in my evening treatments. I try to see myself during my weeks of incapacitation and imagine the stress I would put on my husband to care for it all. I can't see the fairness to him or me, or most especially, my children.

And if I managed to survive through all of it, pregnancy, infancy,

toddling, adolescence, I highly doubt I would make it through the teen years. These are years for the strong and well armored. These years are laced with "attitude". The mental anguish would totally threaten the Zen required to stay healthy. I've lived through them as a pseudo-stepmother and am entering into them as a real stepmother now. All I can say is that I'm grateful for the part-time status.

Had I had the courage and blind faith to just take that plunge, I may have surprised myself and been an awesome mother, but I will never know if there is truth in that. The risk always outweighed the desire. But taking the plunge into stepmotherhood wasn't as scary for me. It was something I thought I might be able to handle and even enjoy. It didn't have the full blown, lifelong commitment attached to it, so it seemed to suit me better. I think I'm a pretty good stepmother. I really adore my stepchildren and I try to respect my role in their lives. It isn't the easiest situation to be thrust into each other's world uninvited, but it is a situation that we all seem to be handling pretty well.

I thought I'd commission the help of my two stepchildren to add some perspective on my abilities (or inadequacies) as a mom, and the impact this disease plays in our family life. When I first met them, David was 11 and Nicole was 8. They are now on the cusp of 15 and 12. They knew nothing about CF before meeting me, and they are now relative experts on its realities.

I asked them a few questions, and they were good sports in humoring me. Here is a sampling of our conversation…

Me: Do you think I'm healthy enough to have a baby?

David: No

Nicole: No…because if you had one, the baby might not be healthy.

Me: Why do you think my disease might interfere with my being a good mother to an infant, an adolescent, and a teenager?

David: Your meds are too time-consuming, since raising an infant and adolescent is a 24/7 job. A teenager would be perfect, since they don't want to be watched.

Me: (Little does he know how much they NEED to be watched!!)

Nicole: You have to do all of your medical stuff and won't have enough time to take care of your kids.

Me: Tell me a couple of reasons why I'm an *awesome* stepmother.

David: You sometimes make good food. You talk. You listen. You know your boundaries.

Nicole: You do fun stuff with me, you make good food, and you're fun to be around.

Me: Here's your chance to let loose and tell me why I'm a *bad* stepmother.

David: Sometimes you talk too much. You aren't around too much.

Me: Your dad and I feel that sometimes it's best I'm not around, because we think you prefer your alone time with him.

David: You shouldn't care what I prefer. You should be a good parent. And a good parent is around.

Me: (Duly noted. Now he'll never get rid of me).

Nicole: You *sometimes* are cranky and mean. You make us late for stuff because you are doing your treatments and we have to wait. You make us do chores.

Me: I'm NEVER cranky and mean.

Adrian: (My husband). Yeah, right.

Me: Do you think my disease gets in the way of family fun sometimes – if so, how?

David: When we go to Uncle Daniel's house to play board games, you never come.

Me: You know that's because I'm allergic to his dog and I'll be in an asthma attack in about 20 minutes.

David: See, you never come.

David: And in New Mexico, we went hiking and you and dad were like miles behind us. You never caught up to us.

Me: Maybe because you and your cousin ran the whole trail?

Nicole: You make us late sometimes. If you have a coughing attack while we are playing a game, then we have to stop and wait.

Me: What do I *EVER* make you late for?

Adrian: EVERYTHING!!

Me: Do you get mad at me ever because of the restrictions of my disease?

David: No

Nicole: No, because you were born like that.

Me: What are some of the things that are unique to someone with CF that you've just grown accustomed to?

David: The treatments and excessive coughing.

Nicole: Pounding, coughing, machines, and sneezing – oh – and *BAD* gas!! (Ha! Ha!).

Me: Very funny.

So there you have it…from the mouths of babes.

I will never regret not having my own children. It would have been an impossible venture for me. But acting as a supporting role in raising someone else's children has been exponentially fulfilling for me and, hopefully, of some benefit to them. I have found my niche, and I have finally realized that, despite all the years of proclaiming it impossible, I might just be a tad bit maternal. ▲

Debra is 47 and has CF. She is a CPA living in Roselle IL with her husband, Adrian, and two Bichon Frise dogs and is stepmother to two part-time live-ins, David and Nicole.

My Experience Being a Parent with CF

By Cara Brahm

My journey as a parent began with the birth of my daughter on April 25, 2001. It brought me into a new area of my life. Any first time parent will tell you there is a lot of "trial and error" involved and lots of learning along the way. My daughter recently turned eight years old and we have come a long way since then. I want to share some background information of my life with CF.

I was diagnosed with CF in middle school, and my two oldest sisters (Lisa and Mindy) were diagnosed then, as well (we come from a family of seven)! I am sure it came as a shock to my parents, but at the same time they were relieved to know what was causing our symptoms.

Anyway, I was very fortunate in my "early CF years" to have few problems. I took my Pulmozyme® and albuterol in my nebulizer every day and was on enzymes, at that time, to help with weight gain. I saw my doctor, usually, just the required every three months and occasionally was put on an oral antibiotic if a problem did arise.

After high school, I went to college and earned an associate degree, entered the workforce, and was married in May of 1998. I continued to live my "normal" life, even though now there were the added responsibilities of being a wife, working outside the home, and maintaining a house.

I found out I was pregnant in the late summer of 2000. We were excited and hoped for everything to be okay. Things went really well throughout the pregnancy and I was followed closely to be sure of a positive outcome.

My daughter arrived with a normal delivery, even though I had to be induced prior. She was a healthy 7 pounds 4¼ ounces. The months following her birth were busy, as could be expected with a new baby. We were fortunate she was a pretty good sleeper at night; lord knows we needed the rest!

In January of 2002, at a scheduled doctor visit, I was told after my usual office "work-ups" that I needed to be placed in the hospital. I could not believe it and was upset by the news. I cried right there in the office. I thought, "I feel fine - I don't need to go to the hospital!" But my numbers told a different story and that was the first of three hospitalizations that year. The next was May and then again in October. Taking care of a new baby had apparently taken a toll on my physical health.

> "I have no regrets having my daughter. In fact, I cannot imagine living without her."

A while after that I again required IVs with hospitalization. In 2005, I received a port and ever since have been able to do "tune ups" at home for myself (my last was a year ago in May). My lung function has decreased in the past few years, but my weight and oxygen levels are good. Thanks to antibiotic therapy and use of *The Vest*® for treatment, I've been pretty stable recently. I try to realize my limitations and have learned to "let things go" until I feel more like doing them.

I think as all of us with CF age, we're going to face obstacles with our health, whether we decide to have children or not. Of course, everyone has to evaluate their own health and if having a child is even a possibility. Talking to your CF team and your family is important before considering whether or not having a child or adoption is right for you.

I have no regrets having my daughter. In fact, I cannot imagine living without her. She is very happy, does great in school, and loves singing. I see the joy she brings to countless others, also. She loves meeting new people and has a special interest in animals.

Last fall, I attended a retreat for persons with serious illnesses. What I remember most is the retreat leader telling us, "Today is all that we have, tomorrow may not come, so we need to live in the now moments." This may sound negative but is true for all of us. Those among us who are blessed with good health do not know what may have to be faced around the corner.

I have learned by having CF to enjoy the days and try not to worry about the future, though I do want to see my daughter grow up into adulthood and be happy. I hope for new treatments and to see a cure for CF in my lifetime. ▲

Cara is 32 and has CF. She does half-time office work. She lives with husband, Ben; daughter, Laurel; and dog, Carma in Lebanon, KY. Her phone number is: 270-692-9143

Rally 'Round CF Roundtable

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Welcome to our first annual fundraising drive!

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- Started in 1990 by adults with CF
- All work is done by volunteers
- We currently have over 1300 subscribers
- No one is turned away for not being able to pay for their subscription
- People in 10 countries receive our newsletter
- Articles from CF Roundtable have been reprinted (with permission) in other countries

Money Facts

- It costs approximately $4.25 to print and mail each copy
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The USACFA Directors thank you for your continued support!

FROM OUR FAMILY PHOTO ALBUM...

CLOCKWISE FROM BOTTOM LEFT: KEVIN (19), JOHN AND MARIA (16), AND JESSICA (14) HELP MOM, JEANIE HANLEY, CELBRATE HER 47TH BIRTHDAY.

LOUIE AND DEBBIE AJINI.

RACHEL MAUGER CELEBRATES HER 26TH BIRTHDAY WITH AN ICE CREAM SUNDAE.

SUSIE BALDWIN AND HER HUSBAND, ADAM LEVY, IN COSTA RICA.

ANDREA EISENMAN AND HER DOG, ERNIE.

STEVE GONZALEZ (BOTTOM) WITH HIS WIFE, BRIDGET, AND SON, DOUGLAS.

CARA, LAUREL AND BEN BRAHM AT LAUREL'S FIRST COMMUNION.

KRISTI AND PAUL FELD WITH DAUGHTER, SARAH, IN THE MIDDLE.

Live Your Destiny

PHOTO BY ALEJANDRA CHAVERRI

Kelsey, David, TJ, McKenna
Jordan, Shannon, Lynnsey, Nina
God sent each child for a reason
Only He knows for what season

What do you mean? Some aren't perfect?
Inherited genetic defect
Their dad and I, we are to blame?
Sheer disbelief and utter shame

Life is not fair! is what I felt
I must live with the hand I'm dealt
The road, so hard, with twist and turn
We each decide what we will learn

Grant me, this day, just what I need
My faith is weak, please intercede!
Believe in God, trust and obey
Just lean on Him, He'll guide the way

Today is glorious, celebrate!
It may not last, don't hesitate
Be of good cheer, please don't despair
He'll keep us in His loving care

So many strive and work and pray
To find a way to save the day
But if it is not meant to be
I must not question Destiny

Heav'n will one day be the cure
There, no more treatments to endure
Imagine life with perfect health
A dream more cherished than great wealth!

The end of sickness and disease
To run and play, do as you please
On lush green grass, on streets of gold
Oh, what a wonder to behold!

Give thanks to God for He is Grand
He knows what we don't understand
The how, the why, the final bell
Fear not, trust God,
love much, live well!

by Kathy Hardy
2002

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

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

DOWNEY *continued from page 1*

delicious looking female than I was in learning about disease.

Long story short, I charmed and wooed and courted and, eighteen months later, a plump priest in a church was asking us to agree to certain terms of perpetual endearment. Andrea Eisenman said, "I do." (didn't I tell you I was lucky?) I found out more about CF along the way. The inhaler. Enzymes. TOBI®. Plugs. Cross-infection. I listen to her breathing while she sleeps, curious and cautious and amazed.

Then, last month, my wife sounded worried. She just got her third sinus infection since meeting me. Her FEV_1 scores had been getting lower. Something was wrong. She wanted me to see an ENT and get my sinuses checked. She knew I had sinus problems, everything from headaches on airplanes to 'Tylenol Sinus' pills on the nightstand. She also knew I didn't like going to the doctor. In my family, one went to the doctor for two things – delivering babies and setting broken bones. Illness was something to be endured. Suffering was good for the soul.

> "In my family, one went to the doctor for two things – delivering babies and setting broken bones."

But my wife is teaching me some things. One is that she knows more about the human body than most people, nurses and doctors included. I'm not very smart, but I'm no fool. I went to see the ENT that week. Two exams, 10 days of Avelox and one CT scan later, there was no doubt that my sinuses were a mess of inflamed tissue and infection. I was a probable source of cross-infection to my wife. I scheduled surgery for the following week.

Surgery was a new thing for me. The way I see it, I'm happy new things are around. I believe they help us progress as a civilization. But I am not interested in experiencing new things. I cross the street when I see them coming. The morning of the scheduled septoplasty, I was in pain–anxiety-induced pain. I remember being wheeled in to the operating room. The strangers in masks. The table that looks like the one they use in Texas for lethal injections. I had sent e-mails to loved ones and strangers alike the night before, hoping to drum up attendance at my funeral, just in case.

It's now the day after. I'm sitting on our couch, a bloody gauze pad taped under my nose. My lovely bride is cooking some matzoh ball soup to help the healing. And maybe I understand a little better the place of cystic fibrosis in our life. My wife asked me to write this article. She even suggested the title, and I'm realizing she's right. This episode IS an example of the "for better, for worse" part of our wedding vows. But it's the "better" part. I got to help my wife stay well. I got pushed outside my comfort zone and I will be stronger for it. We can move forward. We're getting better. ▲

Steve and Andrea were married last September. He learns more about CF every day.

WILLIMAN *continued from page 17*

time in doctor's offices waiting to hear about one of the kids as I did seeing my own docs. But again, no one was ever dealing with something long-term. I guess having CF makes me appreciate that a little more than most parents.

We lived through three kids who needed braces on their teeth, and we taught all of them how to drive. It's always a challenge when you end up stopping two inches from the car in front of you.

There has to be some mention of the sorrow our kids have caused us. Kids are, well, kids, and they sometimes make bad decisions. Ours have made some very bad choices. Nothing breaks my heart more than to see my child choosing to do something that I know (and they know, too, even if they won't admit it) is going to lead to terrible consequences. Some of their choices have resulted in lifetime negative consequences. As mine have become young adults, it's hard to know that there's nothing you can do at such times.

So you tell them you love them and you'll support them emotionally, even though you don't agree with what they've decided. And sometimes they actually come around and return to the values you raised them with. I find it takes a lot of prayer, especially now that they're older.

Yes, for Jon and me, parenting is winding down. Our youngest is 17, going into his senior year of high school. Amy married a great young man several years ago, and we entered a new era with a son-in-law. We also now have a six-month-old grandson, J.J. He has opened a whole different world to us; I love holding his soft little body, only to give him back when it's time for him to go home!

It's been a long road, these 29 years of parenting, full of all sorts of unexpected turns and twists. But I think I'm glad I chose to take it. (Ask me again when the youngest child goes off to college in a year!) ▲

Anne is 56 and has CF. She lives in Middletown, OH.

Pearls of Parenting Wisdom

By Jeanie Hanley, MD

As a parent with CF and three darling teenagers (14, 16 and 19 years old) there are quite a few pearls of wisdom I'd like to think I've picked up about parenting, from my own experiences and those of other parents, and how having CF comes into play. As a pediatrician and allergist, I've interacted with many families, doled out lots of advice and, throughout the years, refined much of that advice, unsolicited and otherwise. Different stages of my children's lives have required continual adjustments in parenting skills. While I can't list all of the pearls that have been learned over my nearly 20 years as a parent and doctor as well as from friends and family, I can point out a few of my favorites.

Some words about parenting in general: First, I believe that most parents, whether or not they have CF, would agree that the early years are the most tiring, sleep-deprived ones, especially with the first child. People advised me to sleep when my child slept. That's very difficult when you've been waiting to perform a million tasks while your child is sleeping. Nonetheless, especially with CF, it is good advice and if you can follow it, you'll be better off because your health will be rewarded. Your health will depend on how much rest you can sneak in.

Second, while every baby's milestones are new and exciting, there are many fears too. It is very easy to lose objectivity when your own child comes down with an illness. Sometimes being in the medical profession helps in diagnosis and treatment, and sometimes being a doctor makes it worse. I often will consider the worst case scenario when my kids become ill beyond the simple cold. Thankfully, a great advantage for a CF parent is our intricate knowledge of nebulizers. With all of our CF equipment, we are ahead of the game when it comes to respiratory illnesses, allowing us to easily treat any coughs that come our child's way.

Third, unlike the early years as a parent, the preadolescent and teenage years involve more time to sleep. One major difference is that, as they grow, the challenges of parenting and their problems also grow. The issues no longer consist simply of whether they can have toy A or toy B that costs 99 cents or kissing a boo-boo, but about "needing" the latest computer or video game, dating, friendships gone awry, bad teachers, grades, the latest cell phones, then losing or damaging same cell phones, and many more issues multiplied by the number of children you have. My philosophy is to be as supportive as possible, be firm about what you believe they want versus need, and above all, keep up my treatments no matter what crisis is occurring.

Speaking of the treatment regimen, there are probably not too many activities I haven't done with my kids while *the Vest*® and/or nebulizers are running – breast-feeding, bottle feeding, burping, food feeding, soothing, changing diapers, playing, laughing, medicating and later, practicing speeches, completing projects, joking, listening, decorating and helping with countless homework assignments. Sometimes, it feels like you're directing an orchestra – saying what to do to get dinner started while proofreading a homework assignment or taking a phone call, knowing that you have to go through the mail, pay bills, contact or phone so-and-so. Despite my husband's very active role in parenting, children require lots of attention and even with both of us giving our undivided attention, it still never seems quite enough.

Another issue that people ask about regarding my kids is how their social life and my having CF affects them. My kids never had to get used to the Vest sessions and nebulizer therapies because it has been a constant in their lives. It's just the way things are. Fortunately, our house is frequently a meeting place for their friends. Although I was worried about their friends' reactions to my constant nebulizing and percussion treatments, it was amazing to witness how accepting just about all of their friends have been. For all these years, I can only think of two instances where one teenage boy who was visiting for the first time was dumbfounded and looked at me as if I was from another planet and the other, a teenage girl, who made a ridiculous comment, the specifics of which I can't remember, but only that the residual ridiculousness of it remains.

Quite the contrary, their teenage friends occasionally sit down and talk with me, especially if they're waiting around for my son or one of my daughters. They've helped me make decorations for upcoming parties, asked questions about exactly what the Vest does or about CF, participated in fundraising events and overall have been terrific teenagers!

Beyond these generalities, the following are some of the "pearls" of wisdom that have worked for my husband and me and seem to work for others. Basically, the happier the

child, the easier your life will be.

Pearl: Always thank and give them positive reinforcement for the little and big things they do. A kid who is appreciated grows up to be a kid who appreciates life.

Pearl: Hold your infant/children as much as you like. I've often heard parents say that they were told not to hold their child too much or else they'd be too dependent on them. Quite the contrary, children who are held frequently, often grow up to be confident children, sure of the love of those around them. On an even more practical, CF-related side, it also builds upper body strength. Definitely a plus in CF.

Pearl: Children are very resilient and adapt very well. In fact, I believe that the treatment routine of a CF parent can be reassuring. They recognize stability in the routine and understand that most often you will be available when you sit down to perform chest percussion and nebulizers.

Last and Probably the Most Important Pearl: If you are hospitalized, it's okay to be selfish. The most difficult times are during hospitalizations. You have to get better. You need rest. I remember my youngest daughter screaming as she left after every visit to the hospital. Although I couldn't help but feel sad as a parent, I knew the best thing for me to do was to stay in the hospital, rest and improve. This behavior stopped by the time she turned six. As long as you have a capable caretaker for your children, you need to let go as much as possible and concentrate on yourself, for once! Returning to the resiliency pearl, they will adapt, you will go home and life will return to whatever normal is for you and your family. ▲

Jeanie is 47 and has CF. She is a physician who lives in Los Angeles with her husband, John, and three teenagers.

EISENMAN *continued from page 1*

had one. And he told me that he did not have green mucus coming from his nose. I thought, "Okay, I will continue to try to figure this out."

As I was stumped, so were my ENT and pulmonologist. I was getting severe sinus infections and needing oral antibiotics frequently, sometimes once a month. And then my PFTs were starting to decline. I was coughing up more mucus, which for me means a few clumps a day, when I had none for many years post-transplant. My pulmonologist, at my insistence, did a bronchoscopy on me, even though he did not believe I had anything to vacuum out. He later reported to me that once he was in there, he found that I was so full of thick, viscous mucus, I never would have been able to cough it out. It was a good thing he did the bronchoscopy. He did not understand why my lungs were so full of that gunk; it did not show up on the x-rays nor did he hear anything unusual when he listened to my chest in the office. I told him my theory of it being my sinuses that were the issue. He thought it was a possibility that it could be draining from my sinuses to my lungs… but why now? Why not several years ago? Good question.

When I saw that my PFTs were deteriorating I consulted my ENT, who is great because he is always interested in my medical theories. I told him what was happening. He thought it over and even though I had just had a minor surgery with him in October 2008, to re-open my left sinus cavity, he was not sure why my sinuses were still giving me problems as much as seven months later.

Then came another sinus infection. And for the life of me, I was mad and I felt it came from nowhere. I should mention that I am a little crazy with compliance to my medical regimen. I steam my sinuses and do nasal lavage two to three times-a-day, inhale TOBI twice-a-day and exercise about six times a week, all while injecting insulin and taking my oral medications, too. There really was no good explanation and I was not near someone with sinus issues. Or was I?

I had a feeling that Steve was somehow inadvertently infecting me and I felt upset and frustrated because he and I have different views about when to see a doctor and when to just wait it out. I don't have the leisure to just let things go. I have to be proactive if something is slightly wrong, because I know how things in post-transplant life can blossom very quickly into a bad infection, etc.

> "I have to be proactive if something is slightly wrong, because I know how things in post-transplant life can blossom very quickly into a bad infection, etc."

After being put on a two week course of antibiotics, I came home that day and asked Steve to go to an ENT. He said he would go to mine. Great. He went to see Dr. Lanny Close, my ENT, at Columbia Presbyterian. Steve explained to him that he did not feel like he had an infection, but had had headaches and pain behind his eyes for as long as he could remember. Dr. Close looked into his sinuses with a scope and declared that he was not sure if he had an infection but if I thought so, he would cover Steve with Avelox

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My Experience as a CF Parent

By Paul Feld

On March 4, 1983, my wife and I adopted, on her first birthday, a little girl we named Sarah. The moment we met, in a solitary room at Catholic Charities in St Louis, Sarah reached out her arms for me to hold her and began to fall down, as she had months to go before she could walk. I grabbed her before she hit the floor, lifted her up to my chest, and we smiled at each other. Renee Zellweger said it best – "You had me at hello.".

That was the easy part. Getting to that place and date took some hard work, some money, and some hard decisions on parenting in general. Not being correctly diagnosed with CF until I was almost 20, I had visions of having a couple of children during my married life. A little while after I was told I had CF, my diagnosing physician explained to me that almost 98% of CF males were sterile due to their tubes being blocked.

My wife and I were still determined to have our own children, so we tried in-vitro fertilization in 1981. It was a relatively new procedure then, and very expensive. We could afford to try it twice. Unfortunately, in both instances, we failed to conceive.

Although very disheartened, and very broke, we went almost another year before bringing up parenting again. We discussed adoption and began to explore the options we had. We could go through the state agency, but we heard the waiting list was long, and it could be five years before a child might be available to us. This was also our least expensive option, and would have provided us more time to save. Our other primary options were to go through a religious-based organization, or attempt foreign adoption. Foreign adoption offered the quickest, yet more risky process, and was by far the most expensive. Our local Catholic Charities organization fell somewhere in between, offering moderate costs and a moderate waiting period. We decided to hook up with Catholic Charities and see where it would lead.

PAUL AND HIS FIRST WIFE, CHRISTINE, WITH SARAH SHORTLY AFTER THEY GOT HER FROM THE ADOPTION AGENCY.

An older lady, named Mary, was our agency contact. She was a 'by the book' counselor, which was both good and bad for us. I guess the only comparison I can make to the adoption process is purchasing your first home. The lender, or agency in this case, wants every bit of personal information they can find on you, your parents, your background, your career, and most importantly, your expected life span. If I recall, they also interview all parents, siblings, grandparents, and in-laws. Obviously, all this is done in the best interest of the child. However, no human being is perfect, and any potential imperfections in a person's actions or personality gets scrutinized to the nth degree.

Having CF played an interesting role in this process as well. On one hand, it was almost proof-positive that we could not conceive our own children, and allowed us to escape some testing that might have been done on us otherwise. On the other hand, the expected life span of a CF adult was mid-20s at the time, and what agency would want to provide a child to a set of parents with one of them expected to die in the next five years?

That became our biggest obstacle for us. I had to convince our counselor of two things: the first that I would do everything in my power to be compliant with my healthcare and never ignore it, and second, if the inevitable did happen earlier than planned, my family and my wife's family would be there to raise the child in a loving home. Our counselor, Mary, gave the OK to move forward with the adoption – with two caveats. We would only be able to adopt one child from their agency, and, while we had hoped and asked to adopt a newborn, we had to modify that request to adopt a child between the age of newborn and two years. We agreed to both, although I would have preferred that my child grow up with a sibling, like I did.

The entire process from beginning to end took about three years in total, and cost us about $3000 (plus

some minor home improvements). I have been able to see Sarah graduate from grade school, high school, and college. She is now 27 and lives in Overland Park, Kansas, while I still live in St Louis. She is independent, and has been 'on her own' since her sophomore year in college. She has brought immediate and long-lasting joy to my life, and continues to do so with her achievements and in overcoming the many obstacles we all face at times in our lives.

Several years ago, through some random act of reading the 'personal' section of the weekend newspaper, I was able to connect Sarah to her birth family (with her permission, of course). Her birth aunt had placed an ad which I just happened to read on Sarah's 19th birthday. I called the number and had two discussions with her aunt, before laying the news on Sarah. She decided, rather quickly, that she'd like to meet her aunt and her birth mother. They connected in person a few weeks later, and she has since developed a relationship with many in her birth family.

While I think the relationship is a positive one for Sarah, I also think it solidified her understanding of why she was put up for adoption and that, in the long run, it was in her best interest. This was something I could never have explained to her based on the information Catholic Charities provided to us.

While the adoption process may have changed somewhat over the last 25 years, I doubt it has changed much. A good agency will always look out for the interest of the child, and I hope this short article can help explain some of the challenges ahead, should you decide to go that route of parenting with CF. ▲

Paul is 52 and has CF. He is a Director of USACFA and is the President. His contact information is on page 2.

and Nasonex spray and would send him for a CT scan in two weeks. Steve came home and recounted the tale. I thought, "Oh, boy, I just made him go to the doctor: a doctor who doesn't take his insurance and who didn't even see anything wrong."

After about a week to ten days, Steve felt a little better. He had stopped sneezing in the middle of the night. He was scheduled for a CT scan and he would then see Dr. Close after that to discuss his situation. He did tell me about a day before the CT scan that he felt it was unnecessary as he was not going to have surgery and that it just invites all sorts of problems. I said, "Just go, you may get to the bottom of what you feel has been an issue for you your whole life." (as it turns out). But part of me felt guilty about making him go to the doctor and worried that if they found nothing, I would have to do some serious apologizing.

No apologies necessary, from me at least. Steve asked me to go to the visit post CT with him. I am glad he asked me to accompany him to see Dr. Close as I did not want to pry and be intrusive. The moment of truth came as Dr. Close put up the x-rays of Steve's sinus cavities. And, lo and behold, his sinuses were full of infection and inflammation. I felt relieved that we had finally gotten to "ground zero". Dr. Close told Steve that he could live his life the way he had been, including the pain and slight congestion, but that because he was married to me, he needed to have the operation or he would keep infecting me and spreading the infections he had.

I have to give kudos to Steve because without even flinching he said, "If it affects Andrea, I will do it ASAP.". I felt guilty about him needing to have the surgery because of me, but Dr. Close also told him that he would not have such bad headaches and would breathe much better. And Steve had really bad headaches. At times they were a combination of a migraine and a rotten sinus headache that he managed by popping handfuls of Tylenol Sinus tabs.

Dr. Close explained the complexity of the surgery and recovery as well as the risks. Which, for a novice like Steve, sounded a bit tough. After the procedure, he would have to steam (with a personal steamer) as much as every hour post-surgery and not blow his nose for many weeks. The surgery, when it was done, about three weeks after that fateful meeting in Dr. Close's office, was really rough on Steve. After the surgery, I spoke with the doctor to see how it went and Dr. Close told me that it was much worse than the CT scan had indicated. He said that it was good that Steve was having this done. Unfortunately, Steve had a lot of pain post surgery for about a day and half. I had had no pain after my two sinus surgeries—but I have been through many surgeries and, who knows, I may have a higher tolerance. I did my best to make him comfortable and did make him some chicken soup—good for almost everything!

I was sorry that Steve had to go through a surgery and suffer. But I also felt validated by my feeling that something was not right and I wanted to get to the bottom of it. Not knowing was making me feel depressed and hopeless. I am learning that marriage, like any relationship, is about sacrifice and communication as well as the fun stuff. I am glad that I was able to communicate to Steve how important it was for me to have him see a doctor, knowing his stance on doing so.

It has been eight weeks since his surgery and he is healing well, and best of all, he is feeling better; which also means, I am feeling better. ▲

Andrea is 44 and has CF. She is a Director of USACFA and is the Executive Editor and Webmaster. Her contact information is on page 2.

Five Lessons for Surviving the Road to Transplant

By Rachel Mauger

I've read many accounts of individuals with CF and/or parents of individuals with CF recounting what it was like dealing with getting the diagnosis of cystic fibrosis. I was diagnosed with CF at birth, due to meconium ileus, so it was my parents who ultimately had to cope with this potentially devastating news more than it was me.

To me, having CF is all I've ever known and is just part of who I am, and I've never thought of it much more differently than the fact that I have blue eyes. My mother made a decision early on to treat me the same as she would any healthy child; hence, I've always done what I had to do to treat the CF, but other than that, I never let CF define me or tell me what I can or cannot do. This mentality has held true even as my treatments have gotten more intensive over the years. I can tell you only from second hand experience how my parents chose to deal with the diagnosis of CF and how it impacted my attitude towards life. I can share with you from my personal, first hand experience what it is like dealing with a diagnosis of end-stage cystic fibrosis and facing the prospect of needing bilateral lung transplant to save one's life.

I've always worked hard to get what I wanted out of life and have been quite successful in this endeavor, while also staying relatively healthy throughout the major part of my life. I grew up with a pretty normal childhood and it wasn't until I was 18 that I had my first real run-in with CF that required a hospital stay and treatment with IV antibiotics. Since then, CF has thrown me a few bumps here and there but I always have dealt with them in stride and, while perhaps causing me a few temporary setbacks in life, it has never stopped me from ultimately going where I wanted to go and doing the things I wanted to do.

By the age of 23, I had just finished my Masters of Education degree in Elementary Math and Science, was soon-to-be married, and was just

RACHEL MAUGER

starting my first real teaching job. I felt like I had my whole life ahead of me and I was finally just getting to where I had worked so hard to be all of my life. My health definitely wasn't what it used to be at this point, but it was still manageable. I felt pretty good and was doing well.

It was at this point in my life that I had my first encounter with the dreaded word "transplant". I had just moved across the state where I was living at the time, so that meant I needed to get established with a new set of doctors. Having been relatively healthy my whole life (for a person with CF) you can imagine my horror when I went to my first appointment with my new CF doctor and, as he reviewed my records and talked to me, he casually mentioned the word transplant in reference to wanting to keep me from needing that for as long as possible.

Prior to this occasion I had never even heard the word transplant used in relation to CF or myself, but I did know enough about transplants in general to know that someone who needed one was dying without it– and surely I wasn't dying! I honestly thought this doctor must be a bit off his rocker as no way was I that sick! He didn't know me and he didn't know my CF and I couldn't help but think how ridiculous it was of him to throw around such a scary word when, if that was ever in my future, it was far off and nothing I needed to start worrying about yet. I had received IV antibiotics only about six times in my life at that point, a small number compared to most CF patients. I had defied CF odds my whole life and done things everyone told me I'd never be able to do, such as competitive gymnastics for 14 years and teaching elementary school; so I wasn't about to let CF or this doctor suddenly start telling me what I now could or could not do.

Over the next year or two that word, "transplant", kept creeping up more and more often as my health seemed to be declining more rapidly, but I continued to keep shaking it off. I still didn't think I really was as sick as they were trying to tell me I was. That was until the spring of last year, when my health suddenly just tanked, requiring two hospital stays within a month of each other and a six week short term disability leave

from teaching. Prior to this I had never been in the hospital more frequently than once-a-year. Although I eventually did recuperate from this exacerbation, I re-stabilized at a much lower breathing capacity baseline. I also had really scared myself and was now faced with accepting the undeniable fact that I was taking on too much. And, for the first time ever in my life, CF was getting in my way. I couldn't outmaneuver it or ignore the reality of its severity any longer - no matter how hard I tried.

While I did go back and finish out teaching the last couple months of that school year, I was slowly becoming more accepting of the reality of where my disease was and what I was realistically able to continue to do and not do. With this reality also came some devastatingly hard decisions, both in and out of my control. In the matter of one summer I went from working full-time at a job I absolutely loved to not working at all and applying for disability. I initiated meeting with and being evaluated by the lung transplant team. My husband, who had previously been very supportive of all of my health issues, suddenly decided that being part of my life and all this was not for him, and I was forced to move back home with my mom after living independently for about eight years. I guess you could say this was me hitting pretty much my rock bottom, to say the least, and I was devastated and depressed. It was as if, overnight, I went from having everything to having nothing.

Well, the good thing about hitting rock bottom is that the only way to go from there is up. I believe everything happens for a reason even if we don't know it at the time, which I can tell you from experience, is frustrating and doesn't make things any easier. However, losing all these things did put into motion a chain of events that has lead me to where I find myself today and is offering me my best chance at a successful transplant and, hence, a second chance at life.

As I embarked on my transplant journey that has led to my recent active listing, I found myself on an emotional roller-coaster that felt like it was spinning out of control. It has taken several months and a lot of lessons learned for me to move on from the original denial I described above, to acceptance and now, ultimately, excitement about transplant and the opportunities that lie ahead for me.

In reflecting on my experience and this transition over the last year, I realized that there are maybe five lessons or key ingredients I've discovered that have allowed me to survive this pre-transplant roller-coaster ride. I'd like to share with you now these five things in the hope that I may be able to help someone else cope with the ups and downs of the pre-transplant stage, just as others who have been through this before me have helped me survive. So, here it goes, five lessons or crucial ingredients needed for surviving the road to transplant:

Lesson #1: Build up your support network. This is, without a doubt, the most important ingredient to surviving all stages of the journey before, during, and after transplant and what has led me to write this article for you today. A strong, multidimensional support network is essential to your well-being on the transplant journey. It should consist of not only your immediate family, closest friends, and transplant team, but also your extended family and all of your friends. It should also include people who have been through what you are going through and those who are currently going through the same thing at the same time.

While I've always had a super strong support network of friends, family, and health care professionals, it was only as I embarked on the journey towards transplant (and was so overwhelmed with everything) that I finally sought out and added this last piece to my support network. I can only wish that I had done this sooner as these people are some of my best sources of support and information, especially during the tough times. The computer was the first avenue I used to reach out to other adults with CF and those experienced with transplant. I joined a couple of online CF listservs and posted on a couple of forums. This is what also led me to *CF Roundtable*. These things have helped me to start regular e-mail correspondences with several individuals that I now think of as members of my primary support team.

Therefore, my advice to anyone beginning the transplant journey is to expand and strengthen your support network beyond just your main support people. Make sure your support people have support people. Get in touch with old friends you've lost touch with over the years and reach out to new friends who have shared and can relate to your experience. Unfortunately, you may find, as I have, that some people who you always thought would be there for you when you needed it most won't be, but I think you'll also discover as I did that there are even more people that you never expected to be there for you who will be.

Lesson #2: Consider seeing some sort of therapist. This could be a counselor, a psychologist, a social worker, a psychiatrist, whomever you are most comfortable with. You need someone to talk to who is not emotionally attached to the situation and to whom you can vent about anything and everything that is going on, without hurting anyone's feelings or constantly being bombarded with very subjective advice that friends and family often unintentionally give. One thing I can guarantee to you as you go

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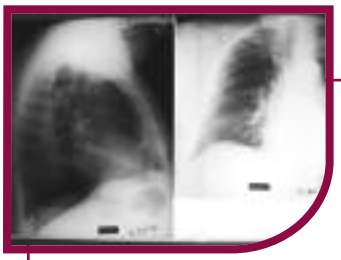

A DEEP BREATH IN

What Is This Column Anyway?

Debbie Ajini

A Deep Breath In is the title of my column that first appeared in the *Autumn 2005* issue of *CF Roundtable*. I am sure the irony of the name is not lost on most of you readers. For those with CF, during PFTs we are asked to take a deep breath in. But it all is a matter of perspective, because my deepest breath may or may not be as deep as yours. But the fact I can still take a breath in is the gift.

If you have read *CF Roundtable* for any length of time you know there are two main components to it; the regular columns like mine, *Speeding Past 50, Information from the Internet, Unplugged,* etc. Almost all of these are written by board members and are intended to be in every issue. And then there are articles that are on just the Focus Topic. These may be by board members but mostly are by other members of our community. For columns like *Speeding Past 50,* the framework is very clear. Kathy is able to speak to focus topics from the perspective of being older. It is up to the author of our regular columns whether they follow the focus topic.

I have been having a period of reflection on this column and its definition - its parameters. Initially when I started this column, I wrote more to my specific issues and how I dealt with them. There was a lot of focus on lists, positive changes, and other more nontraditional means of caring for myself with CF. Then it seems I began writing specifically to the *Focus Topic* for the issue.

I wrote only about issues that I felt I had personal experience with. And I feel every column I have written thus far served a purpose. But I like things to be consistent, neat, organized - so to speak. And I feel my column has been more of a potpourri. Basically I can write about whatever I want, but what is the thread from issue to issue? That is what I am trying to clarify.

I have had a mix of columns including ones that are more fact oriented, like my experience of traveling with oxygen or getting listed for TX. Then there are columns that speak more to the emotional aspects of CF, like "But You Don't Look Sick" or "20 Things You Can Do for 15 Minutes". Is it okay to have both? Of course it is! But for someone who works well with lists and guidelines, I have been feeling a lack of focus for this column.

I am not limiting myself to a box, but I do want to be clearer about the angle I plan to write from. Whether I am writing on the *Focus Topic* or not, I always want it to end on a positive note. Because that is the only way to get through life. I learned that from my mother. No matter what, no matter how many times I get sick or discouraged, there *always* comes the point where I square my shoulders, take *A Deep Breath In* and say, "Enough!". I am in control of how I handle this situation; I still have the power here. I want to give you that power, too, if you don't have it yet. I want those of you who need just that little bit of a push, to read my column and take your deep breath in.

> *"Since, no matter what I write, the fact is simply that I am here to write it. I can take a deep breath in."*

DEBBIE AJINI

And there it is. As I type this, trying to find a framework for my column, it was right there all the time, at the top page of every column I have written. Since, no matter what I write, the fact is simply that I am here to write it. I can take a deep breath in. I will continue to share my struggles and triumphs in appreciating that fact, especially as my deep breath gets harder to do and the possibility of new lungs that breathe very deeply inches closer into my reality.

So while my story may or may not be about the Focus Topic, it will always be about the moments in life that require *A Deep Breath In* and just one step forward. ▲

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

through the transplant journey is that you will experience every emotion in the book. You'll have ups and downs. Sometimes you'll be confident and sure of yourself and feel great, and other times you'll feel awful and wonder if all of this work and stress – to even be eligible for a transplant – is worth it. This person can help you consider things objectively and from points of view you may not have considered. He/she can lend an ear when you just need to vent. He/she can offer strategies to help you deal with all the stressors, from financial to relationship issues, that are most likely going to accompany your life as you journey towards transplant.

I've always had a strong support network, but I was very, very resistant to seeking out this kind of help. All I could think of was all the stereotypes related to people who need to see a "shrink", and if I went it meant I wasn't strong enough to handle things on my own and I was mentally unstable — two qualities I would never use to describe myself. Thankfully, I was pushed by those around me, especially my CF doctor, to look into talk therapy with a clinical psychologist, which felt a little less like the traditional stereotype I had in my head. I came to realize that going didn't mean I was weak or necessarily needed psychotic drugs, although I know these are also sometimes appropriate and effective for some people. This was without a doubt the best "decision" I made and it has been invaluable to my mental and emotional health. It is what has allowed me to handle everything as well as I have while continuing to pursue transplant. Just hearing from a professional that all the thoughts and feelings you're having are completely normal for someone in your given situation is reassuring. While I hope that none of you ever has to face three major life changes at the same time, as I did (loss of: job, husband, and health), I think talking with someone is an invaluable asset that will help you be as prepared as possible - mentally and emotionally - for transplant.

Lesson #3: Educate yourself on life before, during, and after transplant. I'm sure this comes from my background as a teacher, but I truly believe that education and knowledge are power. Over the years I have become an expert on cystic fibrosis, my medications, how to deal with infection, etc. Having a transplant is like trading one disease for another; hence, all my expertise about cystic fibrosis, while far from useless, now must be supplemented with an understanding of transplant and a whole new realm of medications, treatments, restrictions, and responsibilities. Therefore, one of the first things I started doing after seriously beginning to pursue transplant was to get my hands on as much information as I could about the actual transplant process and life after transplant. I've read books and articles about transplant. I've talked with those who recently went through transplant and those 10 years post-transplant. I've researched lung transplantation online. I've also connected with other individuals waiting on the transplant list at my center. By continually expanding your knowledge and understanding of transplant, you will gain confidence in your own ability to deal with transplant as well as be just one more step ahead of the game after transplant, both qualities that will make you that much more likely to have a successful transplant experience.

Lesson #4: Get organized. One of the first things that started happening to me just after I was referred to the transplant team was that I was inundated with paperwork, appointments, and things to do. After going crazy for a little while trying to keep mental checklists of all these things and always hunting for paperwork that easily got misplaced, the perfectionist side of me kicked in and took over and I created "Rachel Mauger's Lung Transplant Binder", complete with a cover sheet for each section and colorfully marked tabs. While I realize this may scream "former teacher", it probably has been the one thing that has helped keep me most sane as I've tried to navigate the pre-transplant road and take care of everything that was asked of me. By just grabbing this binder I can be assured I have everything I need with me for both an emergency medical situation and routine appointments.

While I probably could write a whole article on creating such a binder, I will try to give you a brief overview of the five main sections of my binder. The first section is titled *Appointments* and is where I keep business cards of everyone I've worked with on the transplant team, as well as pages to make notes during appointments and to write questions I want to ask at my next appointment. Any and all papers relating to my appointments with the transplant team and pre-transplant testing also go here. The next section I have is *Insurance* and is for anything I get from my insurance company relating to the transplant process from coverage approval letters to reimbursement forms. Next, and probably the most important and definitely the most used section of my binder that I would encourage each and every prospective transplant candidate to have, is my personal *Medical Info*. On my computer (so that it is easy to revise/update as necessary), I created a detailed list of all of my medications, as well as a sheet outlining my medical history including my diagnosis, medication allergies, previous hospitalizations and surgeries, and my educational/work background. Furthermore, I have a copy of my health care proxy and another document from my computer with names, telephone numbers, and specialties for my entire medical care team as well as contact information for the oxygen company, home-care company, and pharmacies I use. *Directions* is the title of the fourth section and contains written directions to and from the transplant hospital, as well as parking vouchers and now a transportation letter to show local

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TRANSPLANT TALK

Penny Girl

By Colleen Adamson

I have always wanted a dog. I never thought it was possible though; I couldn't even have stuffed animals growing up! I feel about dogs the way most people feel about children – something in my heart just responds in a motherly fashion when I see dogs - but not children. It's not that I hate children; I just do not have a motherly instinct for them. I always thought that was odd since I knew I couldn't have either one, so why wouldn't I long for both? I think that made it obvious that I am not the motherly type when it comes to children, so that made me feel better about not having one. All my other friends couldn't wait to have children, but I am very uncomfortable around them – I don't know what to say to them or how to act. I gravitate toward dogs though!

After my lung transplant, I started thinking about the possibility of having a dog. In the lung transplant literature I read, it didn't say anything about not being able to have a dog; it only said that being near cat litter boxes was a no-no. The possibility of getting a dog swirled around my head for a few years. I asked my lung transplant doctor about having a dog and he said I could! I was so excited! I decided on a name for this nonexistent dog, Penny, because my husband is from Pennsylvania and I am from New York (Pen-NY). I thought it was such a perfect name that I had to have a dog named Penny!

Fate came through for me. Well, actually my in-laws came through for me. My father-in-law got his wife a dog for Christmas three years ago. He researched it for months – which dog doesn't shed and has a nice disposition. He finally decided on a Miniature Schnauzer. They do not shed and they are very sweet. Her name was Nellie; I saw pictures of her and really did not think she was all that cute. When I met her, I thought she was okay but still not that cute. When Nellie was a year old, she had puppies. My father-in-law asked if we wanted a puppy; of course, the answer was yes! My husband, Scott, even was there for the births of the puppies.

We went up a few weeks later to pick out a puppy. Obviously, I wanted a girl to match the name it had already had for years. I sat outside with the puppies for over an hour, trying to decide on a dog. I finally narrowed it down to two dogs; one was pure black and the other was a salt-and-pepper color. My brother-in-law liked the salt-and-pepper one because it was more social. My father-in-law liked the black one for its color. My husband told me it was my decision to make. It was so stressful! Finally, the dogs made the decision for me; the salt-and-pepper one just sat there and looked at me, and the black dog came up and pooped right next to me! Decision made – I chose the salt-and-pepper one. We put her new pink collar on, and she was adorable. I was already in love. I didn't want to leave her at the end of the weekend, and she just looked at us as if to say, "Take me with you!". It broke my heart.

COLLEEN ADAMSON AND PENNY

Penny stayed with my in-laws for ten weeks. My husband went up and got her, and I left work early so I could be there when they arrived. I was so excited! My husband stayed home with her for a week to get her oriented and somewhat trained. She was very sweet and easy to train for the most part. Our next door neighbors came and took her for walks during the day. They loved Penny so much that they also got a puppy. Penny and Rocky grew up together and are great friends. Penny will not go inside if she knows Rocky is outside and vice versa. Penny loves the two children next door also – whenever we say, "The kids", she gets very excited. Just precious!

I am so grateful to be able to have a dog. She has filled a place in my heart that I thought would never be filled. I feel like a mother to her, even though she is a dog. She adores my husband, and when I see them together my heart just melts. I was able to have a dog only because of my lung transplant, and I will be forever grateful to the donor family for not only saving my life but also making it possible for me to be a "mother"▲

Colleen is 40 and has CF. She is a Director of USACFA. Her contact information is on page 2.

DESCH *continued from page 11*

Sometimes, you must move fast. Occasionally, you must move in two directions at the same time. This is not to be taken lightly. Movement is good…ergo…having children is good.

Sometimes a leash helps.

Eating well (I hope)

Okay, this is a stretch…at least for the average American family. For them (but not you) adding a child increases the likelihood of eating fast food, junk food, pizza, goldfish crackers, Kool-aid, Cinnamon Toast Crunch Cereal, etc. (does it sound like I have been there?).

I'm putting this in the positive category because you will have read "legacy" above, and want to be remembered by your children when they are grown and having children of their own as having modeled impeccable nutritional habits.

HELP! (eventually)

There is nothing like raising a child that will speed up the raising of the white flag of "I give up…. I need help!" And this is good. You DO need help. You probably did before children but didn't want to admit it. Now, you are forced to. There is nothing wrong with asking for help. It really does "take a village". In my opinion, it "takes a village" to do a lot of things, but since it is socially acceptable to use this "excuse" when you have a child, use it for all it's worth.

And, of course, the physical challenges:

Sleep

This is the BIG one for the first year or so. Oh, who am I kidding? This is always big. An efficiency for me is when I get my two-hour nap in before noon. But especially during that first year, all I can say is 1) this is NOT the time to split up with your spouse, and 2) a good rule of thumb is that when baby sleeps, you sleep. Seriously. It doesn't matter if you find yourself taking afternoon naps. You will need them. Your immune system needs them. Sleep deprivation is a method of *torture* for a good reason!

Bugs

This is the one everyone warns you about, and I will be no different. Your children will bring home preschool crud, followed by elementary school crud, then middle school crud and, alas, high school crud. Kids are germ dispensers wrapped in cute skin. This is an incontrovertible truth. Denial is not a good defense mechanism for this one. This is one of the most serious physical challenges of parenting… for us.

What can you do? I know some people who have successfully drilled sanitation rules into their children. They wash their hands upon awakening, upon entering the house, and upon every sneeze or cough. No neighborhood child or friend is allowed in the house if they are ill. This is great…and I only wish my kids would play by this rule-book. My saving grace is that divorce has had a silver lining…I **can** send them to their other house when they are sick. Do I? See "Guilt" above.

Time (is time physical?)

I list this one as a "physical" challenge because you *physically* need to get a heck of a lot more done in twenty-four hours than you did before becoming a parent. Sadly, adding a baby to the mix does not remove the need for a disciplined approach to your own health care. In contrast, because of the above challenges, that little creature creates the need for even **more** discipline!

Here's an analogy: Imagine you are jogging slowly to keep ahead of a giant wheel (CF) that is rudely tailing you…always there…always threatening to run you over if you slow down or deviate the slightest from your path (regimen). That baby that you are contemplating creating…that cute little thing that will coo and gurgle so sweetly…is like adding a downward slope, *steep* at times, to your path for the next **EIGHTEEN** years. You have to stay ahead of the game…you have to focus…there is no room for "Oh, I'll just skip that tonight…I'm too tired…"

Energy

Many *CF Roundtable* issues ago I described wellness as the ability to manage energy well. Smoothly balancing energy IN (from eating well, breathing correctly, and taking responsibility and control of your thinking and sensing) with energy OUT (moving, playing/working, communicating, etc.) leads to optimal wellness.

I viscerally remember one day, early after the breakup, when I faced five days of solo childcare for a 3-year-old terror with an understandable adjustment disorder after his world changed abruptly (he's adorably sweet now) and an 18-month-old baby. I lay on my bed, sobbing. All I wanted to do was sleep. I was not just drained physically (I think I was also sick) but, emotionally, I seemingly had nothing left to give. One of my biggest accomplishments in life was getting up that day and just doing it, as they say.

You learn energy management because if you don't…well, it's not pretty. The above scene will not likely occur for you. You will have help. But it is a great example of how energy is a commodity, like time, that has to be managed. And when you are slightly handicapped in that arena by having cystic fibrosis, it only becomes more important to manage it well.

So, now you know everything you need to know to make this decision, right? Wrong. How can you know? You can only make a responsible, informed decision, and then give it your best shot. That's pretty much the case with every decision you face. This is a big one though. You can't send them back. ▲

Julie, is 48 and is a physician who has CF. You may direct your questions regarding CF-related health issues to her at: jdesch@usacfa.org.

UNPLUGGED...
With Steve Gonzalez

By Richard De Nagel

Hello, everyone, and welcome back to another exciting edition of *Unplugged*. I hope everyone is in good health and high spirits. For my part, I am coming off another round of IV antibiotics. Gratefully, all of the nurses, pharmacists and doctors I work with are great. What would we do without the dedicated and loving people who work with CF? All of this brings me to the next person who volunteered to be interviewed for *Unplugged*. He is an old friend with whom I have shared my CF journey. When he e-mailed me a while back to say, "Hi," he didn't know what he was getting himself into.

Steve Gonzalez is his name, and he and I went to the same CF clinic in New York for most of our lives. The date of our first meeting escapes me and I can't remember if it was in the hospital, in the clinic, or at a holiday party years ago. Steve is one of those people who has been in and around my life for a long time and someone who knows me better then many of my closest friends. We have instant rapport and endless conversations, which range from work, to chest PT, to doctors, to personal issues, to family, to enzymes, and back again. It is my pleasure to introduce Steve. Here he is:

1) Name - Steven Gonzalez

2) Age - 44, born 11/22/64

3) Where do you live? Rockville Centre, NY

4) When were you diagnosed with CF?

When I was a little, bitty baby (13 months old).

5) Who is your doctor? Hospital? Do you like him/her?

I have been going to St. Vincent's Hospital and Medical Center, in New York City, since the CF center moved there in the early seventies. Dr. Patricia Walker is my physician and she is terrific. We have a very good relationship, which has developed over the past ten years.

STEVE GONZALEZ

6) How would you describe your health now?

Right now my health is okay. I fluctuate between fair, good, and very good. It's a constant battle that is always there. Thankfully, I have had the ability to bounce back rather quickly when things don't go so well. The older you get, the more time you have to invest in taking care of yourself.

7) What is the newest music in your iPod/CD player?

Believe it or not, I don't own an iPod. As for CDs, I've been listening to a lot of "Snow Patrol", "Kings of Leon", and "The Hold Steady".

8) What is your favorite music in your iPod/CD player?

"The Sharp Things", of course.

9) Are you working? How are you doing with that?

I am working, and have been with the same company for a long time. I work 40+ hours per week.

10) Do you believe in a Higher Power? Are you religious?

I do believe in a higher power. I do go to church a couple times a month. I consider myself more of a spiritual person.

11) What are your hobbies? Does CF interfere?

Sports and music are my passions and both have been part of my life since a very early age. Of course, my number one passion is my music. I am a drummer (as you know) in a very successful band called "The Sharp Things" (see #8). I have been very fortunate to have had the opportunity to perform countless shows throughout the U.S., Canada, and London. As I've gotten older I have to be much more mindful about the care of my body and the impact CF can have if I do too much. I actually listen to my body now (well, most of the time!).

12) What is your relationship status? Happy about that? Does CF interfere?

I have been married for 15 years to a wonderful woman, whom I've

known for the past 22 years. Her name is Bridget. We are very happy together. Does CF interfere in our marriage? Sure, it is very stressful at times for my wife; but, I must say Bridget has been through it all with me and she is a great support. I think when you are in a relationship, and you have a condition (like CF or something else), you really need to take into account how your partner is able to deal with your condition, and the feelings that they experience as well. It can be very overwhelming sometimes.

13) What is your most embarrassing CF moment?

Let's see. Okay, 1985 – coming home from college – I was walking to catch my bus on a crowded, busy street full of people, while calmly having a massive bout of hemoptysis. That was a nice treat.

14) What gets you through the tough days?

Knowing that there will be a tomorrow, and that life is way too short and precious to get overly stressed about things. Also, the fact that I can go home at the end of every day, relax, and enjoy time with my family.

15) What do you hate most about CF?

Everything. Mostly the fact that as I have gotten older, everything can be going along fine and then BAM! CF rears its ugly head, and reminds me that it is still there. It can be a very frustrating thing sometimes.

16) What is your favorite movie? TV show? Why?

I have many favorite movies: "Major League", "Bull Durham", and "Good Will Hunting". I like comedies, and movies that make you think. As for TV, "Two and a Half Men" and anything to do with football.

17) Do you have kids? Want them?

I have a beautiful 8-year-old son named Douglas who is the absolute love of my life. Talk about motivation to stay healthy!! Bridget and I do want another child, so that is in the works. Being a parent is a wonderful thing. There are so many moments that you just treasure and will remember for the rest of your life. Having Douglas has really made me look at my health, and make changes in my daily care, so that I can always be here for him. He is the greatest.

18) What do you look forward to?

A long, happy and healthy life with my family. Recording our fourth record with "The Sharp Things". Being able to play and perform music for the rest of my life.

19) Do you think having CF is a good thing or a bad thing?

Interesting question. Having CF sucks, but you adapt and learn to live with it. I don't let it control my life. It is a part of it. In a way it has made me very strong mentally and it has given me a very good perspective on life, people and what really matters. So to completely answer the question - having CF is not a good thing, but some of the residual effects have been positive.

20) Tell us about your friends?

I have a few close friends whom I have known for over 30 years. They are all very supportive of me, and are there whenever I need them. They understand the CF thing and are cool with it.

21) What is your favorite color?

Blue.

22) Do you spend time with other people who have CF? If so, what do you do, and how important is this to you?

I do not. I do stay in phone/e-mail contact with a few people but outside of that not much. There used to be a support group at my CF center that was very good and I liked that very much. That does not exist any more for obvious reasons, but I do miss that.

23) Do you spend time educating yourself about CF? How important is this to you? What effect does this have on your treatments? Rapport with your doctors? Self-image?

I don't spend much time educating myself about CF, as far as journals and publications. I do love *CF Roundtable* and I speak with my doctor about all the latest info and what is out there. We have a great relationship and talk openly about the many different options and possibilities as I get older. It is important to know certain things, but I don't get crazed about it. I do my treatments faithfully, everyday, and that is the best thing I can do to stay on top of my CF.

As for my self-image...he, he, he, I love me!! No, but seriously, some days I have all the confidence in the world that I can do anything, and some days I look in the mirror, take a step back and do a reality check. But overall I have a very positive self-image.

Steve is one amazing guy! I am lucky to have seen his band, "The Sharper Things", in concert a few times and they are pretty amazing. When I first got to know Steve, during our time in an adult CF support group at St.Vincent's Hospital in New York, I really was just beginning to explore how I felt about having CF and to deal with all the different aspects of the disease. It was the first time I really got to talk with some one else with CF. I heard these amazing people share their CF experiences; all about how they handle work situations, romantic interests, resistance to treatments, cleaning nebs and hating

Continued on page 39

agencies to help expedite my travel to the hospital when I get the call. Last is a section titled *Resources* which contains any source of information I am given by the transplant team or I find on my own that is of value to keep as it gives details about life before, during, and after transplant. I also keep a notepad and calendar in the front pocket of my binder and additionally use this pocket to place papers that need to be added to my binder until I have an opportunity to do so. I even also put a map of the hospital complex on the back cover of my binder and find that it is another frequently used item. While this may sound (and is) time consuming to put together, I assure you that you will thank me later.

Lesson #5: Show your sense of humor. Transplant is a very serious medical procedure that should in no way be taken lightly. However, there is a time and place for everything, and sometimes some of the best ways to deal with the intense seriousness and emotional stress that comes along with transplant is to know when it's appropriate to have a little humor in your life to lighten the mood. I know when I am really down, one of the few things that can most easily and successfully help turn my mood around is joking and laughing with my friends and family. For example, I can't tell you how many stories and smiles developed out of my discovery and first experience using what I like to call the toilet-in-a-cabinet in the ICU room I recently was in, or when my sister did the model strut around my hospital room showing off the stylish yellow isolation gown that staff and visitors have to wear over their clothes. Without these laughs I don't think I would have survived these otherwise very stressful and difficult experiences.

I'm told that "waiting for the call" is the toughest part of the transplant process and let me tell you, three weeks into waiting, as I write these words, I already find it very challenging. I jump every time my phone rings thinking, "This might be it." Nevertheless, it is now that I have no choice but to attempt to continue to live my life as normally as possible while waiting and, armed with these five lessons, be ready to tackle with (hopefully) courage and grace whatever lies ahead for me. With that, I will leave you with one of my current life mottos that both relates to transplant and fully embraces lesson #5: "I'm too sexy for my lungs!"▲

Rachel is 26 and has CF and CFRD. She is listed for bilateral lung transplant at Columbia Medical Center in NYC. She lives in upstate New York and can be contacted at: rstout@kent.edu. You can read a more detailed account of her pre-transplant journey at:www.caringbridge.com/visit/rachelmauger.

Mailbox

Thanks for sharing such great inspiration and knowledge!!!

Stephanie Rath
Brownsburg, IN

Enclosed please find my subscription renewal and a little extra to help defray costs.

I continue to enjoy each and every *CF Roundtable* I receive and it is the only newsletter/magazine I read cover to cover! My husband enjoys it too. Each contributor adds something special to the publication and I feel like I know him or her and sincerely enjoy the insight and experience each person offers. It's nice to know we're all in this together!

I love seeing the birthdays – especially this month's "Milestones", which included a 75-year-old!

Thank you for all your hard work and dedication – it is appreciated.

Dawn McGuinness
Niskayuna, NY

Joe Kowalski would be so proud of the continued excellence of *CF Roundtable* and the commitment of everyone involved in this wonderful publication. Keep up the great work – it is much appreciated and needed by the CF adult community.

Doreen Gagnon
Cheshire, CT

I am an adult with CF. I recently subscribed to *CF Roundtable*. I look forward to being a part of this organization.

Emily Haager
Diamond Bar, CA

Thank you for the great job you do with *CF Roundtable*. It is always informative and helpful.

Marie Roulier
Storrs, CT

Hello *CF Roundtable,* You have a wonderful publication that I have enjoyed for years. I want to thank you for all your work. I am so blessed and so grateful for my life.

Thank you again. God bless you all.

Kathy Harris
Bowie, MD

Thank you for this wonderful publication.

Kim Nunnari
Swampscott, MA

DENAGEL *continued from page 37*

having CF. Before that, I had no idea how much time I spent trying to do everything myself, pushing people away and not talking about this thing that I deal with every day. Our group talked about all of these issues, how to handle them, and more. For the first time in my life I felt part of something and that someone understood me. Interestingly enough, I remember having a difficult time showing up for the group; resistance runs deep.

I miss those people and the important role they played in my life back then. I occasionally get a glimpse of that time, usually when I spend time with or talk to another person with CF. One event that got me all charged up last year was the annual CFRI conference in Redwood City, CA, near my home in San Francisco. The conference is an amazing combination of adults with CF, parents of kids with CF, partners and friends. When it was over I was flying high, and feeling both understood and part of a community. This year's CFRI conference is July 31-August 2. And if you have time, there is a retreat for adults with CF following the conference. It is the only time that people with CF are in the majority!

Until next time … ▲

Rich is 40. He has CF and is a Director of USACFA. His contact information is on page 2.

INTERNET *continued from page 9*

activity. Common side effects of Creon include flatulence (gassiness), abdominal pain, headache, and dizziness. The FDA's Office of Compliance and Office of New Drugs within CDER worked with Creon's manufacturer, Solvay Pharmaceuticals, through the agency's unapproved drugs initiative to help the company come into compliance with federal laws by obtaining FDA approval. All pancreatic enzyme products must obtain FDA approval by April 28, 2010, or be removed from the marketplace. The FDA approved version of Creon will be available later this year.

http://www.fda.gov/bbs/topics/NEWS/2009/NEW02010.html

Antibiotics Could Treat Cystic Fibrosis, Other Genetic Diseases

By modifying the properties of the common antibiotic gentamicin, researchers at the Technion-Israel Institute of Technology have developed what could become an effective treatment for many human genetic diseases, including cystic fibrosis (CF). The findings were published online March 23rd by the Journal of Medicinal chemistry. Gentamicin belongs to a class of antibiotics called aminoglycosides, which are used to treat a wide range of bacterial infections. Studies have shown that gentamicin can counteract genetic diseases, including those mentioned above that occur when mutations cause disruptions of the development processes of proteins.

The drug enables ribosomes (the structures within a cell that carry out protein synthesis) to ignore these disruptions and instead generate healthy, full-length functional proteins. But using gentamicin to treat these diseases requires much higher doses than those commonly prescribed for bacterial infections. At these higher doses gentamicin is non-selective and extremely toxic to humans, with irreversible hearing loss (ototoxicity) being the main negative consequence. In search of a way to bypass these complications, the team led by Professor Timor Baasov of the Technion Faculty of Chemistry modified existing aminoglycoside antibiotic drugs, and carefully monitored biological and toxicity tests of the resulting derivatives. The result is "NB54," a new (and patented) chemical derivative of gentamicin.

http://newswise.com/articles/view/550903/

Tobramycin Inhalation Powder (TIP) Improved Lung Function In Cystic Fibrosis (CF) Patients With Respiratory Pseudomonas Aeruginosa (Pa) Infection

Source: Novartis Pharmaceuticals Corporation

In a Phase III study, TIP, an inhaled investigational formulation of tobramycin, improved lung function (as measured by FEV_1) in cystic fibrosis patients with *Pseudomonas aeruginosa* (Pa) infection, compared to placebo. The data also demonstrated, with respect to secondary endpoints, that TIP decreased sputum Pa density, hospitalization and other antibiotic use in these patients versus placebo. TIP is currently in Phase III development for the management of CF patients with Pa infection. TIP, a dry-powder form of tobramycin, is delivered in approximately 4 to 6 minutes via a hand-held, portable, pocket-sized inhaler device, twice daily.

Adverse events were reported by 75.5% of placebo-treated and 50% of TIP-treated patients. The most commonly reported adverse events with placebo were cough, lung disorders and productive cough. With TIP, the most common adverse events were cough, lung disorders and sore throat. There were no major changes from baseline in vital signs, hematology, blood chemistry or urine protein. Audiology tests at selected sites indicated that there were no clinically meaningful decreases in hearing thresholds. None of the patients reported adverse events related to hearing.

http://www.medicalnewstoday.com/articles/150782.php

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Sugar on Bacteria Surface Serves as a Base for a Web of Resistance

The bacteria responsible for chronic infections in cystic fibrosis patients use one of the sugars on the germs' surface to start building a structure that helps the microbes resist efforts to kill them, new research shows.

Scientists have determined that the bacterial cell-surface sugar, a polysaccharide called Psl, is anchored on the surface of the bacterium as a helix, providing a structure that encourages cell-to-cell interaction. When multiple bacterial cells join together with the help of such a structure, they form what is called a biofilm, a persistent community of bugs that is able to resist the effects of a human immune response, as well as antibiotic drugs. In the case of *Pseudomonas aeruginosa*, the results of biofilm development can be lethal. With Psl clearly identified as the foundation of the matrix, or the architectural glue that holds a biofilm together, scientists hope the research could lead to therapies that target the sugar and thus prevent the development of these masses of bacterial cells.
http://www.newswise.com/articles/view/551473/

Cystic Fibrosis Patients benefit from Orphan Drug Designation

An innovative treatment for infections of the respiratory tract in cystic fibrosis patients has received orphan drug designation in the US. Axentis Pharma of Zurich, Switzerland announced today that this sought-after designation has been granted to its product candidate Fluidosomes-tobramycin, a therapeutic that will soon be tested in Phase II clinical trials. This drug is a liposomal formulation of tobramycin, an innovative treatment for infections of the respiratory tract in patients with cystic fibrosis that is delivered directly to the site of infection via standard nebulizers. Preclinical and Phase I clinical studies support improved safety and efficacy profiles for Fluidosomes-tobramycin as compared to currently marketed treatments for infections of the respiratory tract in patients with cystic fibrosis. The orphan drug designation is granted with respect to treatment of pulmonary infections caused by Pseudomonas aeruginosa. Fluidosomes-tobramycin combines the company's proprietary Fluidosomes technology with the well-established generic drug tobramycin. Utilising synthetic liposomes containing tobramycin, a standard nebulizer delivers the drug directly to the endobronchial sites of infection in cystic fibrosis patients. This may result in prolonged high local drug concentration in the lung, which in turn may lead to higher efficacy and may allow lower doses. Axentis Pharma1s Fluidosome technology uses biocompatible lipids endogenous to the lung that are formulated into small liposomes. In the case of Fluidosome-tobramycin, the interaction between tobramycin and the microbial cell is triggered when the liposomes attach to the outer cell membrane. Tobramycin then leaches into the inner cell compartment, which leads to rapid cell death.
http://tinyurl.com/psmb46

Sodium Channel Blocker Shows Promise as a Potential Treatment for Cystic Fibrosis

Source: American Thoracic Society (ATS)

Cystic fibrosis patients may benefit from a new therapy that increases airway hydration, preventing the buildup of mucous, which is a key factor in the disease, according to researchers at Parion Sciences in Durham, N.C. In normal respiration, the moist surface of the airway allows individuals to effectively clear mucous, keeping airways open and viable. But in individuals with cystic fibrosis, the hydration level of the airway is altered and the airway mucous builds up, interfering with normal respiration. One of the mechanisms causing airways to not clear mucous correctly in these patients involves the body's natural homeostasis of sodium which, when absorbed too quickly from the surface of the airway, causes moisture to become absorbed too quickly. The aerosol-based therapy uses a specific epithelial sodium channel-blocking agent called GS-9411, which prevents sodium from being absorbed across the airway, allowing the surface to remain moist. The increase in moisture allows individuals to more effectively clear the airway of mucous and infectious agents. During the study, researchers applied GS-9411 to airway surface cells grown in the laboratory, and assessed the potency and reversibility of the drug on these cells. Results of the study indicated GS-9411 allowed the cells to retain liquid for more than eight hours. Concurrent animal studies revealed that the agent enhanced mucous clearance for more than four hours.
http://www.newswise.com/articles/view/552217/?sc=mwhn

and

http://healthfinder.gov/news/printnewsstory.aspx?docID=627072

Vertex advances cystic fibrosis candidate

Vertex Pharmaceuticals Inc. has launched Phase 3 clinical trials for a potential treatment for cystic fibrosis. The primary trial for the drug candidate, called VX-770, will be a 48-week trial that is open to patients aged 12 years and older, and two additional trials will be open to patients between the ages of 6 and 11 years. The drug candidate targets a defective protein that is thought to cause the disease, and is designed to improve lung function. The primary trial is seeking to enroll at least 80 patients, and registration is expected to be complete in the first quarter of 2010.

http://tinyurl.com/nshwzm

BACTERIA

Eradication of early Pseudomonas infection in cystic fibrosis. TWR Lee. Chronic Respiratory Disease, Vol. 6, No. 2, 99-107 (2009)

Chronic infection with the environmental bacterium Pseudomonas aeruginosa is associated with greater morbidity and mortality for people with cystic fibrosis. Strict infection control measures including segregation appear to reduce but not eliminate the risk of initial acquisition of the organism. There is now good evidence from randomized controlled trials that early eradication regimens consisting of anti-pseudomonal antibiotics are effective in clearing P. aeruginosa and delaying the development of chronic infection in the majority of subjects. These regimens are safe and cost-effective. Ensuring that such regimens are widely adopted is therefore of considerable importance to improving outcomes for people with cystic fibrosis.
http://crd.sagepub.com/cgi/content/abstract/6/2/99

Microbiological and epidemiological features of clinical respiratory isolates of Burkholderia gladioli.

Christine Segonds, Patricia Clavel-Batut, Michelle Thouverez, Dominique Grenet, Alain Le Coustumier, Patrick Plésiat, and Gérard Chabanon. J. Clin. Microbiol. doi:10.1128/JCM.02489-08

Burkholderia gladioli, primarily known as a plant pathogen, is involved in human infections, especially in patients with cystic fibrosis (CF). In the present study, the first respiratory isolates recovered from 14 CF and 4 non CF French patients, identified via 16S rDNA analysis, were tested for growth on *B. cepacia* selective media, identification by commercial systems, and antimicrobial susceptibility, and were compared using PFGE.

Patients' data was collected. All 18 isolates grew on OFPBL and PCA media, but only 13 grew on BCSA.

API20NE strips did not differentiate *B. gladioli* from *B. cepacia*, whereas Vitek2 GN cards correctly identified 15 isolates. All isolates were susceptible to piperacillin, imipenem, aminoglycosides and ciprofloxacin, and were far less resistant to ticarcillin than *B. cepacia* complex organisms. Fifteen PFGE types were observed among the 18 isolates, but shared types were not identified within epidemiologically related patients. The microbiological follow-up of CF patients showed that colonization was persistent in 3 of 13 documented cases; *B. gladioli* was isolated from post-transplant blood cultures in 1 patient. In non CF patients, *B. gladioli* was associated with intubation (3 cases) or bronchiectasis (1 case). In summary, inclusion of *B. gladioli* in the databases of commercial identification systems should improve diagnosis. In CF patients, this organism is more frequently involved in transient than in chronic infections, but may be responsible for post-transplant complications; patient-to-patient transmission has not been demonstrated to-date. Lastly, *B. gladioli* appears naturally susceptible to aminoglycosides and ciprofloxacin, though resistant isolates may emerge in the course of chronic infections.
http://jcm.asm.org/cgi/content/abstract/JCM.02489-08v1

Mycobacterium abscessus in cystic fibrosis lung transplant recipients: report of 2 cases and risk forrecurrence. Zaidi S, Elidemir O, Heinle JS, McKenzie ED, Schecter MG, Kaplan SL, Dishop MK, Kearney DL, Mallory GB. Transpl Infect Dis. 2009 Mar 9

Mycobacterium abscessus is

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increasingly recognized as an important pathogen in some individuals with advancing lung disease related to cystic fibrosis (CF). Because of its resistance to anti microbial agents and virulence, its presence in the lungs of potential lung transplant recipients can be problematic. We present 2 cases of individuals with CF in whom M. abscessus was present in the preoperative sputum cultures. The organism manifested different degrees of invasiveness in the 2 cases after transplantation with different outcomes, suggesting an approach to future candidates for lung transplantation that may be of clinical significance to their physicians and surgeons.

http://www.ncbi.nlm.nih.gov/pubmed/19298240

Mucoid *Inquilinus limosus* in a young adult with cystic fibrosis. Don Hayes Jr., MD 1, Brian S. Murphy, MD, MPH, Robert J. Kuhn, PharmD, Michael I. Anstead, MD, David J. Feola, PharmD, PhD. Pediatric Pulmonology. Published Online: 11 May 2009

Inquilinus limosus is a -proteobacterium that has been recently isolate in the airways of cystic fibrosis (CF) patients. We report the isolation of a mucoid strain of *I. limosus* from the sputum of a 20-year-old male patient with CF over 1 year that was associated with the clinical, spirometric, and radiographic decline in a previously healthy patient.

http://tinyurl.com/ojkv99

The influence of inhaled corticosteroids and spacer devices on the growth of respiratory pathogenic microorganisms. Tjalling W. de Vries MD, Bart L. Rottier MD, Hylke Visserman, Bob Wilffert PhD and Jan Weel MD, Phd. American Journal of Infection Control. Volume 37, Issue 3, April 2009, Pages 237-240

Guidelines advise weekly cleansing of spacers, with one of the reasons being to prevent the spacers from becoming colonized with respiratory pathogens. Earlier work in clinical settings showed conflicting results. Common respiratory pathogens and *Candida albicans* were applied on Petri dishes with and without inhaled corticosteroids and in 3 brands of spacer devices, with and without inhaled corticosteroids. Growth was measured. After 24 hours, Staphylococcus aureus grew in 7 of 18 spacers (39%); *Pseudomonas aeruginosa* grew in 12 out of 18 spacers (67%); and C albicans survived in 5 of 18 spacers (28%).

Microorganisms survived on Petri dishes with fluticasone and beclomethasone but not when budesonide was applied. One out of 30 metal Nebuhalers (3%) was colonized after 24 hours, whereas of 30 Volumatics 8 (27%) and Aerochambers, 17 (57%) still had viable microorganisms. Application of inhaled steroids did not affect growth in the spacers. The colonization of metal spacers is lower than of spacers made of polycarbonate or polyethylene. C *albicans* can survive in spacers. The survival of microorganisms in spacers is not influenced by inhaled corticosteroids.

http://tinyurl.com/cf8yp4

Studying bacteria in respiratory specimens by using conventional and molecular microbiological approaches. Geraint B Rogers, Thomas T Daniels, Andrew Tuck, Mary P Carroll, Gary J Connett, Gondi JP David and Kenneth D Bruce. BMC *Pulmonary Medicine* 2009, 9:14doi:10.1186/1471-2466-9-14

Drawing from previous studies, the traditional routine diagnostic microbiology evaluation of samples from chronic respiratory conditions may provide an incomplete picture of the bacteria present in airways disease.

Here, the aim was to determine the extent to which routine diagnostic microbiology gave a different assessment of the species present in sputa when analysed by using culture-independent assessment. Due to their focus on isolation of a small group of recognised pathogens, the use of culture-dependent methods to analyse samples from chronic respiratory infections can provide a restricted understanding of the bacterial species present. The use of a culture-independent molecular approach here identifies that there are many bacterial species in samples from CF and COPD patients that may be clinically relevant.

http://www.biomedcentral.com/1471-2466/9/14

TREATMENTS

Unorthodox long-term aerosolized ampicillin use for methicillin-susceptible Staphylococcus aureus lung infection in a cystic fibrosis patient. Luis Máiz, MD, PhD, Adelaida Lamas, MD, PhD, Ana Fernández- Olmos, MD, PhD, Lucrecia Suárez, MD, PhD, Rafael Cantón, MD, PhD. Pediatr Pulmonol. 2009; 44:512-515

Staphylococcus aureus is a significant cause of pulmonary colonization in cystic fibrosis (CF) patients. The optimal strategy of therapy in chronically infected patients with this pathogen is not yet established. We report a successful long-term aerosolized ampicillin treatment of a 14-year-old girl with chronic symptomatic S. aureus lung infection.

http://tinyurl.com/capmny

Efficacy and safety of inhaled aztreonam lysine for airway pseudomonas in cystic fibrosis. Retsch-Bogart GZ, Quittner AL, Gibson RL, Oermann CM, McCoy KS, Montgomery AB, Cooper PJ. Chest. 2009 May;135 (5):1223-32

In this randomized, double-blind, placebo-controlled, international study (AIR-CF1 trial, the short-term efficacy and safety of aztreonam lysine for inhalation (AZLI [an aerosolized monobactam antibiotic]) in patients

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INTERNET *continued from page 42*

with cystic fibrosis (CF) and *Pseudomonas aeruginosa* (PA) airway infection was tested. The primary efficacy end point was change in patient-reported respiratory symptoms (CF-Questionnaire-Revised [CFQ-R] Respiratory Scale). Secondary end points included changes in pulmonary function (FEV_1), sputum PA density, and nonrespiratory CFQ-R scales. Adverse events and minimum inhibitory concentrations of aztreonam for PA were monitored. In patients with CF, PA airway infection, moderate-to-severe lung disease, and no recent use of antipseudomonal antibiotics or azithromycin, 28-day treatment with AZLI significantly improved respiratory symptoms and pulmonary function, and was well tolerated.

http://www.ncbi.nlm.nih.gov/pubmed/19420195?dopt=Abstract ▲

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

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