CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
SUMMER 2015

Documentation I$ Everything

By Janie Davies

My name is Janie Davies. I was diagnosed with CF two years ago at age 66. I am the retired mother of two healthy grown men. I have had chronic lung disease since I was a child. My pulmonary problems were attributed to the whooping cough I had as a very young child. In 1975, at the age of 28, I had a left lower lobectomy. I had a two-year-old and was struggling to breathe. I was advised not to conceive another child because I was high risk. I had my second son 17 months later.

I have been in the hospital more times than I can count. You know the drill, IV antibiotics, PICC lines, midlines, central lines, tunnel central lines and, finally, a port. I've had four documented bouts of pneumococcal pneumonia. I'm told this is very unusual for CF patients. In between all this I had breast cancer and radiation treatments and back surgery, while dealing with exacerbations.

Throughout the years, I had several genetic tests and the results were negative. My diagnosis was non-CF bronchiectasis and chronic sinusitis.

JANIE DAVIES

My pulmonologist was convinced I had CF because of all my symptoms and asked if I had ever had a sweat chloride test. None of my pulmonologists had ever ordered this simple test. I had one and the results were positive. The diagnosis didn't scare me. I finally had some answers. The CF clinic in Dallas had just recently opened. I had been seen by the children's CF doctor at my pulmonologist's office for more than two years. I was promptly transferred to the care of the CF clinic. The treatment plan didn't change much, since I was already on all the medications, but now I had the CF diagnosis.

I had years of case management experience before switching from state to federal government employment. I retired as a Quality Review Analyst. I reviewed sample cases to ensure that the employees were following agency operating procedures and policies. I reviewed their evidentiary documen-

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INSIDE THIS ISSUE

CF ROUNDTABLE
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EDITOR'S NOTES

Welcome to summer. I hope it is a good one where you are. I am sure that we all are happy to see the last of this winter, just finished. Now, if only summer will be kind to all.

There still is time to respond to our survey. We'd like to get your responses. If you have not received a copy of it, send an e-mail to: cfroundtable@usacfa.org and ask for more information about the survey.

We have some changes on the Board of Directors. First, **Paul Feld** has chosen not to run for another term on the board. He served 14 years with time as President and Vice President as well as Chairman of the Fundraising Committee. Thank you, Paul. You will be missed and we wish you well on the next phase of your life.

Two Directors have resigned. We wish **Klyn Elsbury** and **Laurel Avery** well and thank them for their service. We have two new Directors. **Piper Beatty Welsh** is from Denver, CO, and **Reid D'Amico** is from Durham, NC. We look forward to working with them. You can read more about them on page 25.

The Focus of this issue is: What To Expect Post-transplant. **Johanna Libbert** tells of her journey to transplant and the year since. **Jess Newport** relates some of her health troubles after her transplant. **Jennifer Hale** writes of her process of getting listed for new lungs, in "Coughing With A Smile." The "Parenting" column finds **Jamie H.** telling of getting pregnant and having a baby after transplant. **Mark Manginelli** discusses ways to finance transplants in "Protecting What Matters." In "Spirit Medicine," **Isabel Stenzel Byrnes** shares a list of spiritual convictions that have helped her deal with the unknowns of life post-transplant. In "Ask The Attorney," **Beth Sufian** deals with health insurance issues, including transplant costs.

In "Wellness" **Julie Desch** talks about "N of 1" trials. You'll have to read it to know what she is talking about. **Andrea Eisenman** writes about snoring and sleep apnea problems in the "Conversation Corner." Beginning on the first page, **Janie Davies** stresses the importance of documenting everything.

Once again, **Laura Tillman** has compiled a good list of "Information From The Internet." Be sure to check it out. "In The Spotlight" features **Jessika Auger** from Bozeman, MT. **Meranda Honaker** has more information for you in "Clinical Trials." "Speeding Past 50" has some of my thoughts on transplants and life.

Take a look at the future Focus topics that are listed on the opposite page. If there is a topic that interests you, please write about it for us. If you feel you aren't a good writer, don't let that stop you. We are happy to help you with an article. We want to hear from you.

Until next time, stay healthy and happy,
Kathy

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Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Scientists grow "mini-lungs" to aid the study of cystic fibrosis

Researchers at the Wellcome Trust-Medical Research Council Cambridge Stem Cell Institute used skin cells from patients with the most common form of cystic fibrosis caused by a mutation in the CFTR gene referred to as the delta-F508 mutation. They then reprogrammed the skin cells to an induced pluripotent state, the state at which the cells can develop into any type of cell within the body. Using these cells – known as induced pluripotent stem cells, or iPS cells – the researchers were able to re-create embryonic lung development in the lab by activating a process known as gastrulation, in which the cells form distinct layers including the endoderm and then the foregut, from which the lung "grows," and then pushed these cells further to develop into distal airway tissue. The distal airway is the part of the lung responsible for gas exchange.

http://tinyurl.com/ppanrb5

Anthera Announces $3 Million Research Award from Cystic Fibrosis Foundation Therapeutics for Development of Sollpura – a Novel Enzyme Therapy

Anthera Pharmaceuticals, Inc., announced it has received an award from Cystic Fibrosis Foundation Therapeutics Inc. of up to $3 million to support the manufacturing and clinical development of Anthera's novel pancreatic enzyme replacement therapy, Sollpura™ (liprotamase). Liprotamase is an investigational soluble, stable and non-porcine enzyme therapy intended for people with low digestive enzyme levels, or Exocrine Pancreatic Insufficiency (EPI) due to cystic fibrosis and other diseases.

http://tinyurl.com/ooj2ubg

New cystic fibrosis research could help develop treatments to improve muscle function

People suffering from cystic fibrosis have less ability to uptake and use oxygen in their muscles, which leads to exercise intolerance. Researchers have now found that people suffering from CF have limited ability to uptake and use oxygen in their muscles, which gets worse with age. A lack of oxygen leads to a limited ability to exercise, which is an important tool to achieve a better

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LOOKING AHEAD

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

Summer (current) 2015: What To Expect Post-transplant.

Autumn (November) 2015: Incorporating Work Into Our CF Care. (Submissions due September 15, 2015.) Are you working full time? How do you make time for all your treatments and have time for work? Tell us your techniques.

Winter (February) 2016: Dealing With Gastrointestinal Issues. (Submissions due December 15, 2015.) What kinds of gastrointestinal issues have you had? How did you deal with them? What can you tell our readers about how to handle these problems?

Spring (May) 2016: Managing Various Conditions (Under The CF Umbrella.) (Submissions due March 15, 2016.)

ASK THE ATTORNEY

Answers To Readers' Questions

By Beth Sufian, JD

In the past three months readers have asked a variety of questions. People with CF, their family members and members of their healthcare teams can contact the CF Legal Information Hotline at: 1-800-622-0385 or CFLegal@sufianpassamano.com if they have questions about the information below or if they have other questions about the rights of people with CF. All contacts are free and confidential.

1. I have raised money for upcoming transplant-related expenses. I receive SSI benefits and I have Medicaid coverage. Someone told me that the money I have raised may result in me losing my SSI benefits and my Medicaid benefits. Is this true?

A person who receives SSI benefits must meet certain low asset criteria in order to remain eligible for SSI benefits. A single person on SSI cannot have more than a total of $2,000 in assets. A married person cannot have more than $3,000 in assets.

If a person loses SSI benefits, he or she also loses Medicaid benefits. In 23 states the only way for an adult to be eligible for Medicaid is to also be receiving SSI benefits. Children under the age of 18 can be eligible for Medicaid coverage without receiving SSI benefits. Once the child turns 18 in these 23 states, the only way to obtain Medicaid coverage is to also be eligible for SSI benefits. In addition, if the person meets low income criteria and is pregnant the person can be eligible for Medicaid during the pregnancy. In 27 states adults with low assets can obtain Medicaid without being on SSI. However, Medicaid will also have low asset eligibility criteria.

If money raised for transplant-related expenses results in a person having more than the allowable amount of assets for Medicaid eligibility, then the person will not be eligible for Medicaid benefits. The fact that a person will not be able to be listed for transplant without the Medicaid benefits will NOT mean that the asset eligibility criteria will be waived. Many people with CF mistakenly believe that if a person needs a transplant then asset eligibility criteria for SSI and Medicaid will be waived if the person has more assets than are allowed. A person must meet all eligibility criteria for SSI and Medicaid in order to maintain eligibility for benefits.

A person on SSI can put money into a Special Needs Trust. Any money in a Special Needs Trust will not be counted for purpose of determining SSI eligibility. (See article on Special Needs Trust in the Autumn 2014 issue of *CF Roundtable*.)

BETH SUFIAN

There are also Pooled Special Needs Trusts. These are typically funds set up by nonprofit groups. A person can have money donated to an account in her or his name. The money is then part of a Pooled Special Needs Trust. The money is in a Special Needs Trust so it is not counted as an asset for purposes of SSI or Medicaid eligibility. This is different from a Special Needs Trust set up to benefit only one person. Being part of a Pooled Special Needs Trust allows an individual to avoid having to pay an attorney to set up a Special Needs Trust and avoid paying monthly trust fees.

In addition, sometimes money donated to the fund can be a tax deductible donation for the person donating the money. One such fund is the HelpHOPELive fund. More information can be found at: www.helphopelive.org. There are certain restrictions on how funds can be used. Many people with CF have used this group to set up an account for transplant donations, but readers should investigate and determine if the group could help them. No recommendation is being made about this group.

If a person is over the allowable asset amount for SSI or Medicaid eligibility and simply gives the money or assets away to a family member or friend, the person will incur a transfer penalty. The transfer penalty results in a person being ineligible for SSI or Medicaid for five years. Any assets that are over the allowable amount must be used to purchase items the person needs or to pay bills for the person who receives benefits. Receipts for such payments should be kept in the event Social Security or Medicaid requests proof the money was spent and not given away. The person may still be

ineligible in the month the person has assets over the allowable amount.

SSI and Medicaid assets rules are COMPLEX. Please contact the CF Legal Information Hotline for more information on this topic. You should not rely on this general description of SSI and Medicaid assets rules to determine how to deal with assets that are over the allowable amount.

> *"Many people with CF mistakenly believe that if a person needs a transplant then asset eligibility criteria for SSI and Medicaid will be waived if the person has more assets than are allowed."*

A person who receives Social Security Disability Insurance benefits will NOT have any limits on assets in order to be eligible for benefits.

2. I have private health insurance through my husband's employer. Can the insurance company treat me as a person who has Medicare Part B for purposes of determining how much the private insurance company pays on a claim for treatment, even though I did not enroll in Medicare Part B because I was enrolled in a private health insurance plan? My private insurance company says that since I was "eligible" for Medicare Part B they will treat me as having Medicare Part B and therefore they will only pay 20 percent of all Part B claims. My spouse's employer has 50 employees.

A private health insurance plan can treat a policyholder who is eligible for Medicare coverage as a policyholder who actually has Medicare for purposes of determining how much the private insurance company is required to pay on a medical claim for treatment. If a person has Medicare and an employer-based private insurance policy, which policy is considered primary coverage is determined by the number of people employed by the employer. Generally, if there are fewer than 100 employees, then Medicare is primary and the private insurance is secondary. If there are more than 100 employees then the private health insurance policy is primary and Medicare is secondary.

If the employer has fewer than 100 employees then the employer's private health insurance plan would be secondary coverage if a person has Medicare Part B. The employer's private health insurance plan would pay only 20 percent of outpatient services because Medicare would pay 80 percent of the charges at the Medicare rate.

The employer's private health insurance plan can consider a person who is eligible for Medicare benefits as a person who is actually enrolled in Medicare. Very few people with CF know this is possible. A typical scenario is as follows: a person with CF opts out of Medicare Part B because the person does not want to pay the $125 monthly premium since the person has private insurance through a spouse or parent who is employed. The person with CF does not realize that the private health insurance plan has a clause in the policy that treats a person who is eligible for Medicare benefits as a person who actually has Medicare benefits.

Many problems with coverage of medical treatment follow. If the person did not have Medicare at the time the treatment charges were incurred and the private insurance plan will pay only 20 percent of charges, then the person with CF is responsible for 80 percent of the charges that would have been paid by Medicare if the person had been enrolled in Medicare. For example, a person who is post-transplant has a sinus CT scan and the charge is $4,000. The person did not enroll in Medicare Part B but the private insurance plan through their spouse, whose employer has 50 employees, treats a person who is eligible for Medicare as a person with Medicare. The private plan pays 20 percent of the charges, $800. The person with CF then is required to pay 80 percent of the charges, which is $3,200. Since the person did not actually enroll in Medicare, there is no way to have Medicare pay the charges.

It is important to check your private health insurance policy to see if it treats a person who is eligible for Medicare benefits as a person who actually has Medicare, for purposes of determining how much the private health insurance plan pays on claims for medical treatment.

In addition, there are certain private health insurance plans that exclude coverage entirely for anyone either enrolled in Medicare or who is eligible for Medicare coverage. The private insurance company is allowed to have such a clause in its policy.

3. My employer is refusing to give me paperwork that I need to request intermittent leave under the Family Medical Leave Act. I had a transplant two years ago and now I am experiencing rejection. I need to undergo treatment for rejection but think I can still work three days a week. I am eligible for FMLA leave, but I know I

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SPIRIT MEDICINE

"I AM..." Spiritual Convictions

By Isabel Stenzel Byrnes

The topic of this *CF Roundtable* is "what to expect post-transplant." The word "expect" is loaded. Sometimes, by expecting something, we increase our suffering when we do not get what we expect. To me, expectation is a form of grasping for control. Before my transplant, my motto was, "Expect the worst, hope for the best." In a quirky way, this phrase eased my fear of dying on the operating table, or having some calamity occur after transplant.

After a lifetime of worsening CF, including a few traumatic experiences and witnessing CF peers not do well, my mind was conditioned to think that lung transplant would result in poor outcomes. Dr. Rick Hanson, author of *Buddha's Brain*, writes, "Most fears are exaggerated. As you go through life, your brain acquires expectations based on your experiences, particularly negative ones. When situations occur that are even remotely similar, your brain automatically applies its expectations to them; if it expects pain or loss, or even just the threat of these, it pulses fear signals. But because of the negativity bias of the brain, many expectations of pain or loss are overstated or completely unfounded." So expecting a bad outcome was purely based on fear. The good news is that my motto also included a conviction of hope. I had to believe that if more transplants were failures, they wouldn't be doing them so routinely; insurance wouldn't be covering them, and they'd still be experimental. I had to prepare myself to struggle or even die, to harness and mobilize my strength and also to cultivate hope to quench the fear. The bottom line was that expectations can be healthy when they offer possibilities of both positive and negative outcomes.

In this "Spirit Medicine" article, rather than focus on expectations, I'd like to switch gears and share a list of my spiritual convictions that have helped me deal with the unknowns of life post-transplant. Convictions are positive statements of intent or mantras that we live by. They are broad enough to encompass how I lived with CF before transplant as well. Whether I lived or died, whether I was healthy or sick, these convictions would always be true. Convictions are spiritually important because they expand our view from the concrete realities of our lives to something wider. They help us feel grounded in what's most important, and they keep us inspired. They challenge any previously conditioned negative thought pattern. They fuel our will to live—which is what we need most when living with CF—pre- or post-transplant.

ISABEL STENZEL BYRNES

So here are my convictions. They start with the most affirming statement, "I am." I am and always will be me... no matter what happens to this body of mine. And no matter what my expectations are.

I am hungry.

I don't just mean the prednisone-induced hunger. Yes, in the beginning I wanted to eat everything in sight! And that is a metaphorical statement, too. With a new life, I wanted to consume everything available to me. After transplant, my life force was given a turbo-boost, and I was thrown into opportunities available to most healthy people. I could now exercise without coughing, I could plan trips and fulfill those plans. I could focus my attention not only on personal health needs, but on reading, learning and focusing on professional interests. I could dream, set goals, aim for things and live out my fullest potential. I could explore who I am outside of my CF identity. I get to make up for all my lost opportunities in my pre-transplant days. Now 11 years out, I am still not satiated.

I am "NOOGTT."

After transplant, I realized that

my life wasn't all about me anymore. After fighting all my life against the end, my donor died unexpectedly. His family endured what I had feared for my family all along...they grieved a devastating and tragic separation. It was now up to me to honor him by living with and for him. I also realized that my family, friends and healthcare providers witnessed my resurrection and were profoundly changed. My CF peers got to see the potential of life for them. Similarly, I looked up to my CF peers who were 10, 15, 20 years post-transplant, feeling awe and disbelief that perhaps I could live that long. I also saw peers who never came out of surgery, who died within a few months, who suffered with multiple problems post-transplant and then died. I have spent much of my post-transplant life mindful of strong fluctuations in my emotions: fear, anxiety, dread, hope, ecstasy, joy. A spiritual practice that helps me during times of uncertainty can be best described as "nougat" or NOOGTT. I'm **N**ot the **O**nly **O**ne **G**oing **T**hrough **T**his. My struggles aren't unique, and I don't have to feel alone. When difficult feelings arise, I know others have experienced them too. So I don't have to take them so seriously for myself.

> *"The bottom line was that expectations can be healthy when they offer possibilities of both positive and negative outcomes."*

I am hopeful.

CF demands a life of hope amidst seemingly hopeless odds. In my post-transplant life, I have tried to sustain positive expectancy. The statistics about lung transplants haven't changed much in 10 years. I hoped for five years, but accepted that even if I got to breathe easy for a few months, I'd be content. I never, ever imagined that I'd be with my husband longer post-transplant than pre-transplant. This freedom from CF has allowed us to each pursue our own interests and grow professionally and interdependently. This is beyond my imagination. As for the future—I remain hopeful. I could still succumb to cancer, rejection, infection...or I could struggle with normal conditions of aging. I am open to all possibilities.

I am surprised.

I once just wanted to breathe. Now every time I look around the corner, there's something new and exciting to experience. Most of it is good; some of it is hard. But there's a newness to everything that keeps me interested, focused, alert. When I got a renewal application for my driver's license, I was surprised to still be here. When I do a PFT, I'm surprised by the numbers. A sense of surprise cultivates a "beginner's mind"—a spiritual concept in meditation that lets me examine all my experiences with curiosity and wonder.

I am maturing.

Getting older allows me to grow in relationship with God and Spirit. I also feel more in touch with who I am. I have been given the chance to enter the "normal" world and reduce my self-consciousness about being different. I have been given time to gain self-awareness. I did not receive this gift of life only to enjoy life. I received this life extension in order to experience life in its entirety. And so that means fully living—losing, hurting, enduring, dreading, mourning along with loving, learning, gaining and growing. All of these experiences guide my maturity.

I am surrendering.

Living with a vulnerable body invites surrender. I know I have to be obsessively compliant to stay healthy (with meds, exercise, sun protection etc.), but I also know anything could go wrong at any time. I've seen that with friends and with my sister. Much of my outcome is out of my control. It is in the hands of doctors, fate, God's will, luck, preordination or the unknown. My time is so limited. Everything I am able to do—bagpiping, swimming, hiking—is temporary. What this does is keep me humble; to practice letting go and accepting that these joys are FOR NOW. I surrender by placing my hopes into prayer and by relinquishing control to God. Then I know I am doing everything possible to live as long as I can, and the rest is up to God.

I am grateful.

When I woke from transplant, I was blown away that all the stars had to be aligned for me to receive this gift of lungs. The organ procurement organization staff, the donor family and their hospital team, surgeons, the helicopter pilot, my medical team all had to coordinate for a smooth, efficient process to transplant the most viable lungs into my rapidly declining body. It is difficult to fathom the magnitude of gratitude that a transplant recipient can feel unless you are one. I live day-by-day full of astonishment and amazement at the fortune I've been blessed with. This gratitude helps buffer the ongoing stressors that I endure as a transplant recipient. Gratitude is the foundation of my spiritual practice—it connects me to God more than anything else.

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SPEEDING PAST 50...

Living The Life That Is

By Kathy Russell

What a summer we are having, and it's only June! We had almost no winter and our spring was much drier than normal. While all of this mild weather makes it quite pleasant in the short run, it bodes ill for the long haul. We are in a drought and starting into our driest months. I hope we don't end up with water shortages. (I have great empathy for everyone in California who is suffering with limited water. We experienced that, when we lived in southern California in the 1970s, and I know that it is no fun.) The rest of the country still is experiencing odd weather phenomena. Too cold in the winter, too wet or too hot the rest of the time... and oh, those winds! I hope we'll get out of this weather cycle soon.

I have seen TV specials on global warming and the melting of glaciers. Now there is concern about the warming of the permafrost in Alaska. That would cause the release of thousands of tons of carbon dioxide into the air, which would not be good for humans. I hope that we (humans) can do something to save our planet. Then maybe we wouldn't have to put up with so many El Niños or La Niñas.

I am old enough to have experienced many unusual weather cycles. I understand that this may be just another part of life. I still do not necessarily like it. In the same way, I do not always like the changes in my health. I understand that they will occur, but I don't have to like it. As my lung function goes down, I have to learn new ways to navigate life. I have to rest more often, and I have to limit how much exertion I experience. I really don't like limitations.

Recently we went to visit the Museum of Flight at Boeing Field in Seattle, WA. I have been there before and enjoyed it a lot. I was looking forward to going again. We talked about how I could manage the exertion, and I agreed that I would use a wheelchair. This is a big deal to me. I always have been able to do things for myself. That no longer is true...and I hate it. I like being independent.

I am so fortunate to have Paul, my husband, who looks after me with such care. He lifts my portable oxygen concentrator (POC) in and out of the car, carries it up any hills and lifts it into my shopping carts. The POC isn't very heavy, but I don't have to use my limited energy to hoist it around. He also helps me change batteries, when that becomes necessary. I really appreciate the help with that, since I tend to get a little panicky when the O_2 has to be turned off, even though it is for only a few minutes. My lungs are greedy for the oxygen.

He helps me in many other ways. I do the cooking, because I still enjoy cooking, and he does the clean-up. That is a real luxury to me. It is so nice to be able to get up from the table and not have to be concerned about clearing it and cleaning up the dishes.

KATHY RUSSELL

We grocery shop together and he does the lifting for me. He also bags the groceries and loads them into the car. When we get home, he carries them into the kitchen and helps me unload them. Another chore he has taken over is making the bed, which has gotten to be way too much for me to do. He gathers the laundry on wash days and helps me sort it. Then he carries it to the washer and may even load the washer for me. He also moves the damp laundry from the washer to the dryer, which saves me a lot of energy. Of course, if I am feeling ill, he does the whole shebang. He even folds or hangs everything. If I am really lucky and feeling very well, he will take the washed laundry out to the solar dryer so that I can hang it out in the sun. I love the way it smells and feels after being dried in the great outdoors. I am happy to use some energy to get this special result.

I mentioned that my lungs are greedy for oxygen—the Focus topic of this issue deals with lung transplants. That is something that I will never experience. Fortunately, my lungs still are sustaining me. It's a good thing that they are, because I doubt that anyone would transplant lungs into a person past age 71!

I get a lot of support from my friends who have had transplants. They all have gone through the problems that I am experiencing, and they can give me good advice. It is reassuring to be able to talk about my status with them. I know they understand what I am feeling, and they can give me suggestions of how to deal with each problem, as it arises.

I don't know if I could accept a transplant if I were a viable candidate. I am a pragmatist and believe that I have to play with the cards that I was dealt. It would be "going against type" for me to have a transplant. I think that if I were young and needed a transplant, I might feel differently about it. We'll never know how I would react, so this is just speculation on my part.

I am delighted when each of my friends who receives lungs heals and does well. I am so happy that they were able to get good lungs, and I want them to live long and happy lives with as few problems as possible. I ache in sympathy for them when they have post-transplant problems. Some have had repeat transplants and others have had to have other organs, such as kidneys, transplanted. Then there are the battles with various cancers. Some have had thyroid cancer, others have dealt with skin cancers and others have fought still other cancers such as sinus cancer and melanoma, after transplants. I understand why it happens, but I still feel pain for them when it does. Having had cancer, I understand what a blow it can be to get that diagnosis. I was so fortunate, when I had cancer, because I have a very healthy immune system and felt very little effect of having cancer. Few are so lucky.

Another thing that my post-transplant friends understand, that is harder for others to get, is that it is very difficult to talk on the phone for more than a few minutes. I think it may be because I have to use more air to be understood on the phone. After a short conversation, I start to cough and need to stop talking. (Some people are hurt when I want to stop a call before they are ready to. Such is life.) On the days that I wake up in the morning ready to take a nap, they understand. On the days when my O_2 saturations are such that my brain is fuzzy, they understand. When I am unable to complete a task I have volunteered to do, they understand. They know about being too tired to care about anything. They also know about wanting to do things but just not having enough energy to do them. They know of the frustration that losing lung function brings. This, too, is life.

I remember one friend talking about how she had never planned for what she would do after transplant. It was as if she hadn't expected to survive. She was really surprised when she woke up after surgery and was *alive!* It just hadn't occurred to her that she might survive. She adjusted quite quickly and made the most of her new life.

Others had put off doing things until "after" the surgery. Many friends have said that it is important to live the life that is, rather than waiting for the life that might never come. I agree with that.

Still, I yearn for things that I haven't been able to do for quite a while. I used to love sitting at my sewing machine for hours to finish a garment or other project. I haven't done much sewing in years. My back was giving me such trouble that the thought of sitting up for any extended period of time just didn't interest me. I hope that will change and that I will be able to get back to sewing my own clothes. I really enjoy making something that fits and is comfortable. My back is giving me some better days, so maybe I'll be able to sew soon.

My friends who have had transplants find that they are able to do things that they haven't done for years or maybe had never been able to do. Some hike or swim or run or bike as they never could in their previous lives. Others travel to exotic places or places that are not so exotic but had been out of reach to them. What a treat! I applaud them.

Several of my friends have participated in the Transplant Olympics and have done well. Hooray for them! Can you imagine running or swimming in an Olympic setting? How about playing tennis at an Olympic level? More power to them! Or how about playing bagpipes at the opening ceremony? One of my friends did that! I applaud her.

One thing that I know for sure is that all of my friends who have received donor organs are extremely grateful to the donor families, and they do their best to honor their donors. I am so grateful to each donor and each donor family, as well. Without them and their generous gifts, my friends would not have survived as long as they have. (One of my friends survived for more than 22 years after transplant.) What a wonderful legacy for each donor.

I think that living life as it comes is important, but you must be ready to deal with the unexpected things, too. Stay flexible enough to accept a miracle if it should present itself to you. If you need a transplant, I hope that good organs become available to you, quickly. ▲

Kathy is 71 and has CF. She is the Managing Editor of CF Roundtable. *Her contact information is on page 2.*

In Memory

Gina Rocchetti, 46
Quaker Hill, CT
Died December 14, 2014

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

CF Roundtable, PO Box 1618, Gresham, OR 97030-0519.
E-mail to:
cfroundtable@usacfa.org

COUGHING WITH A SMILE...

While I Breathe I Have Hope

Dum Spiro Spero "While I breathe, I hope."

By Jennifer Hale

I will never forget the words coming out of my doctor's mouth back in February 2015. I was there for a regular CF clinic appointment and knew my health was bad and had been bad for several years now. I fought back from mycobacterium abscessus, but M. abscessus and living with a progressive disease has taken its toll. My doctor said, "It is time to start the listing process and get listed for a double lung transplant."

I am very grateful my husband was with me that day. You see, he has been coming with me now to CF clinic. Every time I go to clinic these last few years it has been nothing but decline in lung function and to me that is bad news. Well, this time the news was that I needed a double lung transplant. As I heard these words being spoken and transplant coordinators starting to enter the room, I just started to cry. What a thing to hear when you're not expecting it at that moment. When you are thinking to yourself, "I am gonna come back from all this. I am going to increase my lung function. I am going to get off oxygen and I am going to beat this!" I have always beaten CF. I have been fighting for 43 years and I was not gonna stop now. As the coordinators came strolling in and Kleenex boxes were being passed around to me, the harsh reality of what was going to lie ahead of me was scary, daunting, unknown and defeating.

My lung function has been nosediving pretty badly for three years now and I am now at 25 percent. I am 43 years old living with a progressive disease and need new lungs. Growing up, I never thought I would be somebody who was going to need a transplant. It wasn't even on my radar. I have been compliant my whole life and active my whole life. Transplant was not going to happen to me. I never even thought about it. Until now. Now it is at the forefront of my mind—waiting for that phone to ring. Waiting for my second chance. Waiting for my miracle.

The first step in this journey is a three-day evaluation. It is three full days of tests starting from early morning to late in the day. Nothing hurts—it is just an exhausting experience. There is a lot of waiting around. A lot of walking all over the hospital. However, I did use a wheelchair part of the day. I used it mainly due to the fact that my blood sugar was low—I was fasting for a test and the procedure was at the other end of the hospital. If I had walked I would have bottomed out for sure. So I happily accepted being pushed in the wheelchair, giving my body a chance to rest and lower my oxygen needs.

> *"Nobody knows when his or her time is up. That is why it is so important to just live for each day."*

JENNIFER HALE

I got through the evaluation and then it was time for the report to go to a Medical Review Board for review. I got denied. Wait...what? The concern was the mycobacterium I had a couple years back. Is it really gone because if not it can cause major issues and deathly consequences if it is still in my system. I was told it would eat through healthy lungs. So now I was dealing with the possibility of not being able to get a transplant. A transplant that was going save my life. An idea that I was still trying to wrap my head around and that I really needed right now. Here I am fighting in my head going back and forth in my brain thinking: "Do I really need this now? I can live in this state of health, right? I am not THAT bad, right?" As I am playing the game in my head trying to accept my fate, I am now being told I might not even have the chance to come to terms with needing a transplant.

The next step was a sputum culture tested at the DNA level to see if the mycobacterium abscessus was truly gone. Praise GOD, the test came back negative! No mycobacterium.

The Medical Review Board went over my case again and I am now approved for transplant. Doctors feel confident the M. abscessus does not lurk in my body anymore. However, it is not absolute and there will be special protocol put in place for when I go through the transplant surgery to wash out my lung cavity of any bacteria.

"The emotional roller coaster that has been going on since being told I needed a transplant has had its ups and downs."

This was now in the month of March/April of 2015 and I needed to get some vaccines before I could get on the actual transplant list. (You cannot have a live vaccine in your body when transplanted.) Now I had to wait the recommended time period for the vaccines to "wear off" (so to speak) to get listed. I am happy to say I am listed as of June 9, 2015!!!

The emotional roller coaster that has been going on since being told I needed a transplant has had its ups and downs. It is so scary to know you need a transplant. I wrestled a lot with trying to believe it truly was time. Which is kind of crazy because I was on oxygen and my lung function was struggling to stay in the low 30s. But I just kept thinking I can live in this state of health. I'd rather struggle and live. At least I will still be living rather than take the chance of transplant and I could die. But what I was denying to myself was the fact that I am dying. I cannot live in the state of health that I am in for many more years. But I can live for many, many years with new lungs. I have a chance with new lungs. I have no chance with my current lungs. But it is hard to see that through the emotions and tears, because transplant is scary. It can have grave consequences. But as I say that, the funny part is, at the rate I am going I have grave consequences. I do not have a chance right now to live many, many years. But I do have a chance with a double lung transplant. But it has taken me a few months to see the light.

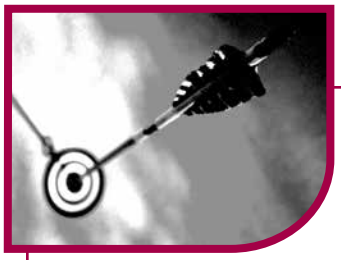
The minute I was told I needed a transplant, I created a spiral notebook. I decorated it with uplifting quotes. Imagine that, quotes. If you have followed my column, I am the queen of quotes and they really help me lift my spirits. The biggest fear I have had is being scared of not having enough time. Not living long. It has been very hard to imagine that at 43 years old this could be it. I am not ready to go. What has helped me manage this fear is a quote I have come up with myself. I just keep saying to myself, "It is about each day, the years will take care of themselves." Look, nobody knows when his or her time is up. That is why it is so important to just live for each day. To be thankful that you have this moment, this time, this day. I feel it also takes a lot of pressure off when you're only focusing on the now. Tomorrow will take care of itself and it will come. But all I can do right now is live in the moment. Cherish the moments I have with my husband, family and friends. When you really, I mean really, embrace and accept living for the present, it truly takes away the fear and pressure of what the future holds.

Another quote that I have come up with that is helping me is, "While there are options, there is hope. Where there is hope, there is life." The option of being able to be a recipient of new lungs is phenomenal. I am actually going to have the chance to live many years. To live breathing! I do not know what it is like to breathe. To breathe like a "healthy" person. It is kind of exciting that I will be able to feel what it is like to breathe! I am very much looking forward to that. It is funny how involuntary breathing is. I think we take it for granted. I never noticed my breathing when showering. Or getting dressed or making my bed. Now, each movement, each activity triggers me to be out of breath. Activities that I have not even thought twice about now have me out of breath. I am amazed how this can be. But my new lungs are going to allow me to breathe again. To breathe better than I have ever breathed in my life. Just the thought of that is a miracle. A miracle I will live and witness for myself. A miracle that a donor family will give me. What a gift!

As I wait for the phone to ring. I continue to work through my emotions of what this journey is going to be for me. At times I get very, very scared. But at other times I am a little excited. And through this journey I have had family and friends supporting me every step of the way and I cannot thank them enough. How do you say thank you for all the love and support during such an emotional time?

Lastly, I dedicate this to my husband. For if not for him, I would not fight this hard. I would not make it through this journey. He is my light in the dark, he is my EVERYTHING. I dedicate this final quote to him, "To the world you may be but one, but to one you are the world." Mark, you are my world. You are my life. Thank you for loving me! I can truly say I have lived a complete life because I had you! My husband, my love. ▲

Jennifer is 43 and has CF. She and her husband, Mark, live in St. Petersburg, FL. You may reach her at: jhale@usacfa.org.



FOCUS TOPIC

WHAT TO EXPECT POST-TRANSPLANT

Jo's Second Wind

By Johanna Libbert

Despite respiratory and digestive symptoms, my CF was misdiagnosed for years until I was nine years old in 1975. Life took a drastic turn with medicines, treatments, chest-tapping and a special diet. Back then, kids with CF were encouraged to decrease their fat intake.

My parents took great care of me and I was hospitalized very few times as a child. The hospitalizations increased after high school. I managed to get through college and become a Registered Respiratory Therapist. I worked at a hospital for seven years. I was full-time, then part-time and eventually quit due to frequent infections and fatigue. I started receiving Social Security Disability Insurance a few years later.

My health slowly declined. In 2005, my CF pulmonologists encouraged me to be evaluated for lung transplant, since my FEV_1 was in the low 30s. They recommended Barnes Jewish Hospital in St. Louis, MO, which is three-and-a-half hours away from my home in Richland, IN.

The rules for lung transplant were changing. It was no longer necessary to get listed years in advance and wait your turn; the most seriously ill climbed to the top quickly. I wasn't on oxygen yet and had good and bad days, so I decided to postpone transplant.

The next time I was evaluated was in 2009. I had been needing oxygen at night for a couple of years and needed antibiotics more often. My FEV_1 was 29 percent, but I still wasn't ready.

Even though I wanted to have a transplant, it's hard to know when to get listed. When I was sick, I was ready, but when I felt a little better, I'd want to wait. I needed to get as much as possible out of my old lungs, because there is such a short life expectancy on donor lungs.

In 2011, my oxygen needs increased: I needed it with exercise. My FEV_1 actually was better – at 33 percent. My $CO_2$was 45, but my PO_2 was worse. Even though I was having more bad than good days, I decided it was still premature.

JOHANNA LIBBERT IN MAY 2014 BEFORE TRANSPLANT (LEFT) AND MAY 2015 AFTER TRANSPLANT (RIGHT).

The early summer of 2012, my sweet Labrador, Ebony, passed away. She was the last of our pets and had become my best friend. We had *slow* in common; she was slow and old and I was slow and sick. I could not stay well and was on continuous oxygen. I was worried about her if I had a transplant and had to be gone for three months afterward for rehab. God took that worry away. I was devastated at her sudden death for a while but slowly came to the realization that I *was* ready for transplant.

In December 2012, we went back to St. Louis. My FEV_1 was 22 percent and I had blood gases of CO_2 at 56 and PO_2 at 45. I completed all four days of testing, which included a heart catheterization. The doctors agreed that I should be listed. So, after taking pre- and post-transplant classes and affirming that I wasn't allergic to Cefepime (the antibiotic that Barnes uses after transplant), I was officially listed in February 2013.

I was CMV (cytomegalovirus) -negative and wanted a CMV-negative donor to eliminate the chance of CMV infection after transplant. Only half of the population is CMV-negative, so my donor pool was smaller.

We packed the trunk camper shell with everything for a three-month stay in St. Louis afterward. We were pre-

pared when we received "the call" to head for St. Louis, no matter what we were doing.

After a year on the list, I was needing more O_2. My CO_2 kept creeping up so I was put on bipap to keep it down. Bipap also provided some relief when my lungs were especially hard to ventilate. They were so stiff and heavy that at times I wondered if I would stop breathing. I had violent coughing fits to expel lots of foul mucus day and night. I took eight nebulizer treatments a day with CPT; my vest machine became ineffective by that point so my husband tapped me by hand. I used all my strength to cough and breathe. My arms gave out after several PICC lines so I had a port placed in my chest for IV antibiotics every other month. There were no more good days, and I could never stay well. I didn't want to go anywhere or do anything so my husband lost his companion.

After a year on the list, I switched my status to accept any donor. Meanwhile, many caring family and friends held benefits for us along with a lot of prayers. They put money in a special account called "Jo's Second Wind." In the fall, a year and a half later, I thought transplant wasn't going to happen: Maybe God had a different plan for me. On September 30, 2014, we went to the transplant clinic in St. Louis for a check-up. Back home at midnight, Barnes called to say they had a donor!

On October 1, 2014, at 48 years old, I received two healthy, pink lungs. My old lungs were dark brown, swollen and disfigured with big red blebs on the surface. They started surgery at 9 a.m. and it lasted five hours. The lungs were from a young lady in the tristate area and were very good. She was CMV-negative and nearly a perfect match.

I was off the ventilator within 24 hours. I was nervous about waking up on the ventilator but I don't remember anything except the extubation. The first thing I noticed was my pink finger and toe nails and how easy it was to breathe. My lungs felt so light. My biggest problem was getting choked taking pills or eating meat for about a month. A bronchoscopy showed some aspiration but the other one was fine.

Two days after transplant, I went from ICU to step-down care. I was on one liter of oxygen and had one chest tube removed. I slept all night without coughing once. The next day I walked for the first time. My lungs did really well, but I had bad back pain from the surgery and moderate to severe scoliosis. Physical therapy and my family helped me walk with my chest tubes, IV pole, oxygen and a wheelchair – in case I couldn't make it back. Five days after surgery, I moved to a regular room. I walked 1,000 feet but had to take four sit-down breaks because of my back pain.

The next morning, our truck was burglarized in the hospital parking garage. In the cab, they had stolen my traveling compressor and my jacket. In the camper shell, I had packed a lot of my favorite clothes in several suitcases. Friends had given me an expensive bag with my name embroidered on it, and church members had made a quilt. I had packed lots of things to entertain myself; books, DVDs, CDs and player, sewing supplies, 150 quilt blocks etc. There were tote bags with medication for three months, toiletries, makeup, cleaning supplies etc., all gone. It was an emotional setback, but I focused on the incredible gift I had received and the wonderful opportunities that lay ahead.

My dear husband, Phil, dealt with the theft, reports and insurance. It was so great having him stay with me in the hospital. He helped me to the bathroom countless times. He was a pro at getting my chest tube unit and oxygen attached to the IV pole. He was a big comfort to me. That day, they took out two more chest tubes so only one more was left!

The next morning, I woke up at the end of my bed. I saw the chest tube and thought I'd been in a car accident. When the nurse came in, she gave me a strange look and asked, "Are you all right? Do you know where you are?" No to both. She told me I was in the hospital and had had a lung transplant. Everything started coming back to me, but for a while I couldn't remember anything. It was a strange and scary feeling. Later that day, the nurse weaned me off oxygen for good. I walked my 1,000-feet goal and started exercising on a portable treadmill in my room.

October 11, they removed the last chest tube from my left lung—and it collapsed. I couldn't tell because my breathing was so good. It was such a relief to get the chest tubes out; they were painful and uncomfortable.

When I was discharged, two weeks after my surgery on October 14, 2014, we stayed at a duplex ten minutes from the hospital. Phil moved us in and took care of getting my medications lined out and loading my pillbox (no small feat!). When my parents came to stay with me during the week, he made sure we knew where to go for rehab and appointments before he went back to work. I am forever grateful to Barnes and my family and friends for their excellent care.

On October 20, the stitches were removed from my chest tube incisions so I could shower. The transplant incisions were just glued together. The water felt strange on my skin because my stomach, thighs and breasts were numb. It was good that my stomach was numb so I didn't feel all those heparin shots!

My schedule involved daily rehab sessions with the treadmill and leg weights

Continued on page 14

at the hospital. Chest x-ray and labs twice a week and doctor's appointment once a week. I wore a mask everywhere to keep from getting sick since the anti-rejection drugs decrease my immunity.

On October 23, I had a bronchoscopy with biopsies to check for infection and rejection; neither was found. I could stop the Cayston aerosol I had been taking since leaving the hospital. It was great not to take treatments, clean equipment, CPT, oxygen or bipap. I could sleep all night and laugh without coughing. Phil was sick when he came on the weekend and coughed a lot at night; what a switch! For years he endured all my coughing in bed and never complained.

After transplant, I coughed occasionally from sinus drainage, getting choked or after a bronchoscopy. There was a squeezing sensation around my chest under my breasts like I had a really tight underwire bra on. It would hurt when I coughed but not as much as when I sneezed. Before transplant, I'd take several breaths to get a sneeze going and sometimes it would just fizzle out. Afterward, I still tried to sneeze the same way even though I had enough air to sneeze right. The same was true for singing. I had to learn how to use my new lungs and not breathe the way I did with my old ones.

On November 3, the pneumothorax was resolved so I could start arm exercises and PFTs. My first FEV was 90 percent, a far cry from 22 percent! I went gung ho on arm exercises all week. My stomach and chest got so tight and painful, my appetite waned and I dropped to 110 pounds (from 115 pounds). I didn't feel like myself; I was very emotional and cried a lot. I missed home and Phil. I had strange notions; I never thought of killing myself but when we watched a crime story, I was jealous of the dead victim because her problems were over. I had never felt that way before. The doctor prescribed an antidepressant and it helped. The best medicine, however, was when I went home for Thanksgiving break. It was so good for my spirit, and I went back to St. Louis in a better mood. I continued with daily rehab going from 0.6 mph to 2.8 mph on the treadmill. I continued arm and leg weights but with softer gusto.

There is a lovely proverb that is so appropriate for my life: Just when the caterpillar thought the world was over, it became a butterfly. I was ready to spread my wings and soar.

On December 20, 2014, I went home for good. My FEV was 117 percent and I weighed 118 pounds.

I wrote a letter to my donor's family and received their letter in January 2015. Natalie was an athletic 17-year-old girl who died from injuries sustained in a single-car crash. She was described as a caring and generous person and was on the cusp of beginning her journey to an independent adult life. She was adventurous and we shared some of the same interests (zip lining, traveling and roller coaster riding). She participated in the dance team at her high school. I felt sad and guilty because *she* should be using her wonderful lungs. But I am forever grateful for her family's generosity at the worst time of their lives. Jesus is my savior and Natalie is my hero.

> "I was off the ventilator within 24 hours. I was nervous about waking up on the ventilator but I don't remember anything except the extubation."

I am seven months post-transplant. My six-month visit involved more tests than a typical monthly checkup. My bronchoscopy revealed no infection or rejection. It is amazing that I haven't been sick since transplant. The anti-rejection medicines are causing some problems: a little high blood pressure, cholesterol and fluid retention. My osteoporosis is worse so I take a bone pill every week. My diabetes is a little worse so along with my Humalog injections with meals I take Humulin in the morning as well. My immunity is down so I take immunoglobulin infusions monthly so I'll have to keep my port. My iron level was low so I had eight iron infusions. I take 42 pills a day.

I don't have any big life-altering ambitions. It's just really great to enjoy the little things in life again: walking in our woods, driving a car, singing in church, cleaning, cooking, shopping, and being able to help my family and friends instead of them always helping me. My husband and I like to travel and now I can do things with him again. It will definitely be different not packing all the machines. I am looking forward to family outings this summer, and I walked in the CF Foundation Walk instead of riding in a wheelchair with oxygen like the past two years.

Right now I'm not planning on getting a paid job; I definitely wouldn't go back to respiratory therapy because of the infection factor. I have filed to stop my Social Security Disability. We will be on a tighter budget, but it's great that I'm too healthy to qualify. My transplant experience was painful at times and a lot of hard work for many people but definitely worth it. So many people throughout my life have helped me survive. I am so grateful.

I adhere to a daily exercise routine including treadmill and weights. My FEV is 127 percent. I weigh 139 pounds because I can eat a lot without getting short of breath and I don't burn

all my calories up coughing and breathing. Dairy products no longer cause congestion in my lungs. The numbness left after a couple of months except my breasts. I am just now starting to get feeling back in them. I still have the sensation of a tight band encircling my ribs under my breasts.

I've coughed up only a fleck of mucus twice since transplant. Before, I was a mucus factory; it's unbelievable the amount of mucus a body can produce at such a fast rate. Most days, I don't even think about all the work we had to do every day just to keep me breathing. It's strange how easy it is to forget unpleasant things.

I hope people with CF won't have to rely on transplants to further their lives. The thought of transplant is scary because the outcome is unknown and the anti-rejection drugs have unpleasant side effects.

It's best to treat each day as a gift; that's why it's called the present! May everyone have health and happiness and, if need be, a second wind. ▲

Johanna is 48 and has CF. She lives in Richland, IN.

POETRY CORNER

EBONY, JOHANNA LIBBERT'S DOG.

My Best Friend'

By Johanna Libbert

Comfort and peace were her gifts to me;
her little sweet self.
She hung on my every word and I loved her thoughtful silence;
adoring companions.
Thankfulness and praise in a lovely forest paradise;
our earthly Heaven.
Content with our simple existence, I didn't realize
my grave situation; my slow demise.
So heartbroken by her sudden death; deep despair.
A shattered life has no need for a lung transplant;
no will to live.
Daily prayer and time helped to soothe the pain;
brighter days ahead.
Her death was not my undoing but an awakening;
my epiphany.
Beautiful new lungs will herald a life of healthy freedom;
my second wind.
A daily struggle to breathe, I pray it's not much longer;
so sick and weary.
Remembering my heavenly girl brings a smile;
Her little sweet self.

It's Always Darkest Before The Dawn

By Jess Newport

> "I learned that transplant is not just a light switch, flicking from sick to healthy. There is so much gray area."

My misadventures in getting my bilateral lung transplant five years ago had its fair share of vast swamps, troll bridges and treacherous cliffs. After waiting seven months at my initial transplant center, we were told that Florida was having a "lung drought" and they recommended me to Duke.

Leaving my sister to watch the home in Tampa, my parents, dog and I drove 666 miles feeling like the time bomb inside me was ticking louder than ever. Duke had an overwhelming program, including rehab five days a week, but what other choice did I have?

While I had waited in Florida, I had continued to drive but was lucky if I walked 500 feet in a day. I could not imagine walking laps pushing my oxygen in front of me like it was a baby I was caring for, because it was. Living with lungs that functioned only about 12 percent, I was not really living. I was too busy trying to breathe, jumping through the doctors' hoops and just putting one foot in front of the other in an effort to prove that I deserved to live.

I had a pretty typical recovery after my surgery. My new lungs continued to blow better every day. My stomach, on the other hand, gave me the most trouble. I wrote an article for *CF Roundtable* in 2013, called "Gastro-WHAT? A Journey After Transplant." Even after a Nissen fundoplication, I had such debilitating nausea that I was questioning why I'd thought transplant was for me. No medication helped the nausea and doctors seemed to think it was in my head.

Random stomach pain led to getting my gallbladder removed that Christmas. Here I was in the hospital, at Christmas, eight months after my transplant, and I still was not well. This is when I learned that transplant is not just a light switch, flicking from sick to healthy. There is so much gray area. It is as though that switch was on a dimmer.

That winter and spring were the toughest times in my life, second only to the time prior to my surgery. My depression was the defining factor. When the nausea and pain didn't keep me in bed, I was isolating myself and trying desperately to become motivated. I had two college degrees to finish and plenty of pleasurable things I wanted to do.

I remember that May forming a "Jess's Breathe Team" for Great Strides.

JEREMY LAMBERT AND JESS NEWPORT.

The day of the event, I had gotten myself so stressed out, that I ended up vomiting in the back of my car at the walk and had to leave to go to the ER. The bottom line was that I knew I wasn't using my new lungs to the best of my ability, and that made me feel guilty. This was all a vicious cycle until I finally got my diagnosis of "chronic pancreatitis" and was put on new medication.

At last, things were looking up. I graduated college in May 2012. The desire to finish my degrees also motivated me to continue my education. When I finally moved out of my parents' house to Durham after two very difficult years post-transplant, it felt like a new chapter had begun. I've been living in Durham for two-and-a-half years now and it feels like home. Healthwise, the only struggles I face are from the effects of the immunosuppression drugs.

I'm the happiest I ever remember being! I have found things that make me happy, and I made them a priority. For one, I volunteer at my local library, because of my love of books. I've found workouts I enjoy, such as yoga and dragon boat paddling, and these attack my depression and anxiety better than medication.

It has taken me four years since my lung transplant to feel well. I am a partly-recycled, better version of myself. I know that unfortunate times will come again, but I feel like I have the foundation to keep my light shining bright. ▲

Jess is 28 and has CF. She lives in Durham, NC.

DAVIES continued from page 1

tation to support their decisions. One very important skill I learned was to document my cases. Setting follow-up dates on my PC or calendar was essential to keeping up with my cases.

I use my case management experience to keep up with Medicare and my private health insurance and multiple hospital visits per year, medications and everything else that comes with CF. I document everything. I use a notebook to keep a daily log of the date and time I take each medication (inhaled and oral) and telephone conversations with doctors, suppliers, distributors and insurance company. I know it might seem tedious to document the time and date of tests and medications administered during a hospital stay. I also have a small notebook where I jot down names of the nurses, residents, interns, respiratory therapists and any other hospital personnel who enter my room.

For example, my last hospitalization I jotted down the time I was wheeling into my room: 7:13 p.m. Nurse T took my vitals and I was introduced to the Tech. I jotted down the blood pressure (BP), temperature (Temp), heart rate (HR) and oxygen (O_2)saturation. 8:21 p.m. RT Xopenex and 7 percent HyperSal Sodium Chloride. 8:53 p.m. Voriconozole. 8:55 p.m. Merrum IV started. 10:18 p.m. Cipro IV started. 10:50 p.m. Tech took vitals. Again, I jotted down the BP, Temp, HR and O_2 saturation. I might be feeling sick and tired, but it takes only a few seconds to jot down the information. Now I know when my next IV antibiotic will start, and I can track my vitals. Sure it's in their computers, but I don't have access to that information. I utilize this information to privately rate the staff when I get discharged. I can write my thank-you notes and mention specific names and their good work or any complaints.

Another reason I keep documentation is that I have been billed for medications and tests I never received. I make it a practice to request an itemized bill from the hospital. Once I review the bill, I request a nurse review. In the past, my hospital bill has been adjusted by thousands of dollars.

In addition to documentation, it pays to know your insurance and policy. My doctor prescribed the Kitabis Pak. Unfortunately, the distributor, IV Solutions in Lubbock, ran the script under my private prescription plan. I requested they run it though Medicare first. Medicare pays 80 percent for two CF medications under durable medical equipment and my private insurance pays 20 percent. The medications are nebulizer inhalation solution Pulmozyme and generic tobramycin. I received a call in April that Medicare approved my Kitabis Pak prescription. Without the CF diagnosis, I would be paying over $2,000 prescription co-pay for a 30-day supply. I thought other readers would like to know, since the distributors are not aware of this.

I think the Kitabis Pak is fantastic since it comes with the PARI LC Plus nebulizer handset. They will also be furnishing the DeVilbiss Pulmo-Aide air compressor at no cost to me. It's much like the Altera eRapid Nebulizer system that was furnished with Cayston. Medicare will pay for only one PARI LC Plus every six months. I guess they don't consider the fact that CF patients have several nebulized medications and require three or four handsets in one sitting.

I'm glad I have case management experience to keep up with medications and everything else that comes with CF. I believe you are your own advocate. If there is one piece of advice I would give to the readers it is to document everything. ▲

Janie is 68 and has CF. She is retired from the SSA. She lives in Arlington, TX, and is divorced. She is the mother of two healthy adult sons and grandmother to three. She loves to dance, read and travel. You may contact her at: Jdavies0116@att.net

YOU CANNOT FAIL

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

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

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

SUFIAN continued from page 5

need to complete certain paperwork to obtain approval for the leave. What do I do?

There is no requirement in the law that a certain form be used to request FMLA leave. The law does require certain pieces of information be included in a request for leave.

If an employer refuses to provide FMLA paperwork, an employee still can make a request in writing for FMLA leave. The person could explain in the letter that the employer was refusing to provide the employee with FMLA paperwork but that there is an immediate need for leave. There should always be a discussion of why an employee needs FMLA leave included in any request for leave. A letter from the treating physician explaining why the leave is needed is important.

Often employers do not like giving employees intermittent leave and may try to put obstacles in the employee's way so that intermittent leave is not requested. An employee whose rights have been violated under the FMLA can file a complaint with the Department of Labor.

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: 1-800-622-0385. The Hotline provides free and confidential legal information to people with CF, their families and their CF Care Center teams. The Hotline is supported by funding from the CF Foundation. Callers speak to an attorney employed by the Hotline. The Hotline is the only service that provides free legal information from attorneys who focus their practice on the rights of people with CF. The Hotline can also be reached by e-mail at: CFLegal@sufianpassamano.com ▲

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

PHOTO BY ROB ROHDE

A Needy Girl

She needs me.
Her body heat emanates to soothe me.
Those deep dark brown eyes gaze at me lovingly and heal me.
I talk to her, telling her how much I love her
When I lie with her, I am completely safe
There are no worries ahead
Everything will be okay
When I am apart, fear overcomes me
Who else will love me?
A fragile composition
An unknown future
We connect and save each other
We met at the shelter when we were both frail
We needed to walk to get stronger
We remember the tubes, the sounds of machines, the pills
The hospital smells
Days before the end was inevitable,
Fate and a miracle brought us both back from the dead
Only hidden scars reflect how close we came.
She walked me,
Short walks at first, and then longer
Now, we abound with energy
Escapes to romp in the snow, hike high into the mountains.
Spoiled by life, we indulge in feeling good, cuddling in softness
These are times to celebrate
The love we share for life and each other.
I've almost lost you once
Now I want to live as long as you
Your life expectancy matches mine
Now, we share a hidden fear
Who will live longer?
I can't handle you leaving me.
I need you.

-I. Stenzel Byrnes, 2006

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

FROM OUR FAMILY PHOTO ALBUM...

JENNIFER AND MARK HALE.

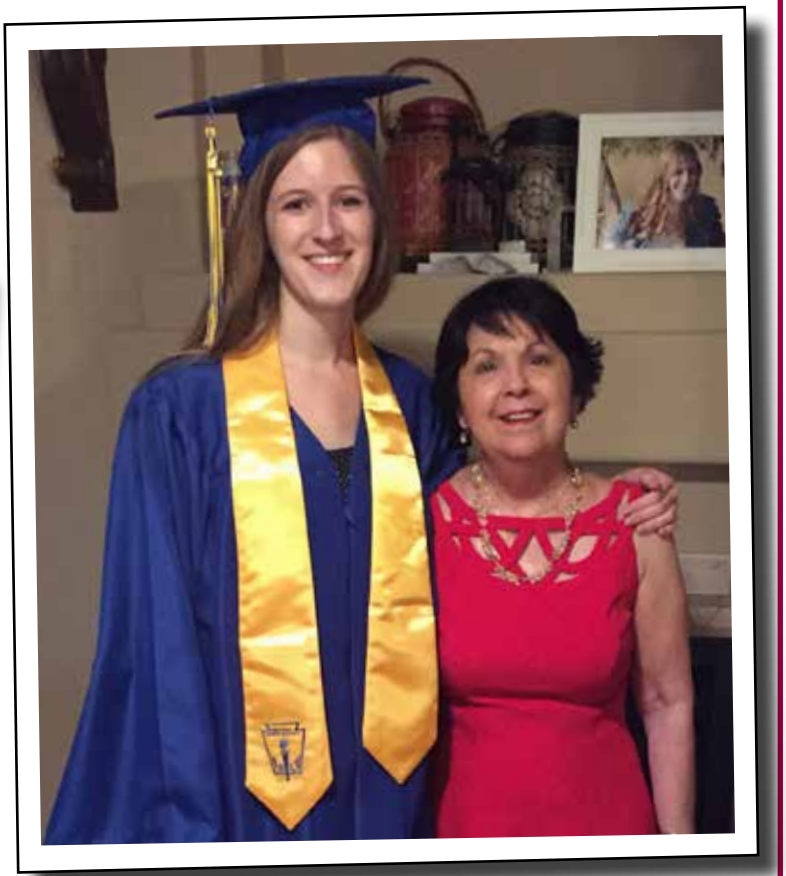
JAMIE H. HOLDING ELAINE ROSE AT 2 MONTHS AND 2 DAYS OLD.

THANKSGIVING 2014 IN HILTON HEAD ISLAND, SC. FROM LEFT TO RIGHT: REID D'AMICO, HIS MOTHER DONNA DELIA D'AMICO, KHAKI (DOG), HIS BROTHER SCOTT D'AMICO, AND HIS FATHER DAVID D'AMICO.

JESSIKA AUGER AT MONTANA STATE UNIVERSITY CATAPALOOZA 2014.

PIPER BEATTY WELSH AT GREATER NY WALK FOR THE GREAT STRIDES IN 2015 WITH HER 6-MONTH-OLD NEPHEW, LASZLO BEATTY SIDON.

ANDREA EISENMAN AND SCRUFFY.



JANIE DAVIES WITH HER GRANDDAUGHTER, HENLEY BRYANT. HENLEY GRADUATED FROM SAHUARITA HIGH SCHOOL IN ARIZONA MAY 20, 2015.

IN THE SPOTLIGHT
With Jessika Auger

By Jeanie Hanley and Andrea Eisenman

Jess played in her first Roller Derby bout, "Game of Throws," in May 2015. Having joined the team on a whim, she became a fast learner and despite all the challenges of school, CF and a recent hospitalization she rebounded and was able to compete in this very physical sport. She even received Bozeman, Montana's Teddy Award from the local TV sports reporter for her grit and perseverance that week. Prior to her first bout, she had been hospitalized and rumor has it that she was practicing skating with her IV pole in tow. Roller Derby aside, Jess is a young adult who tackles CF just as she tackles an opponent. She does what she can but has also learned not to miss out on the fun.

Jessika is 21 years old, grew up in Livingston, MT, and now lives in Bozeman, where she leads a life full of spunk. She snowboards, hikes, rides her motorcycle, fundraises for CF and much more. As a junior at Montana State University (MSU), she is getting better at balancing her social life, managing CF and maintaining her independence. She is a force to be reckoned with and is a fabulous reminder to us all to live in the moment and tackle what life has to offer. Please welcome our newest star. Spotlight, please!

At what age were you diagnosed? I was diagnosed at eight years of age. I was sick frequently as a child, but when my mother took me to see the same doctor, he repeatedly said my mom was crazy and there was nothing wrong with her child. Eventually, he kicked her out of his office insisting nothing was wrong. Finally, we went to another pediatrician and she immediately knew something was wrong. I had pneumonia for quite a few weeks and once that was cleared up she recommended a specialist four hours from home. I was diagnosed then, and they immediately tested my six-year-old brother, who turned out to be positive for CF as well.

JESSIKA AUGER

What was everyone's reaction? I had no idea what I was in for, and it took me a while to fully understand what having CF meant. My mother took action immediately and started figuring out what we needed medication-wise. I think she was relieved a little to finally be able to get me medicine that stopped me from being sick all the time.

How was the transition to a new CF doctor? I had hospital admissions in Denver until the middle of high school, although clinic visits were in Billings. Now I do both clinic and hospital admissions in Billings, which is only two hours from home instead of 12! It was nice being able to be close to home, and my new doctor communicated a lot with the team still in Denver, so the transition was not as hard as it could have been.

What areas are affected by CF? When I was younger, I mainly had lung problems. Now that I am older, I have problems in my sinuses with frequent congestion, a pancreas on the verge of having CFRD, and digestion issues. I occasionally experience joint pain, mainly in my lower back and knees, which sometimes hinders my ability to exercise.

How did you decide on the college? I have an amazing support system with my family, but I also wanted a little independence. MSU is a 40-minute drive from my home, so I am close enough where I can call if I need anything and someone would be able to come get me if needed, but also I am able to live close to campus in an apartment.

What is your major? I am majoring in early childhood education and minoring in psychology. I want to do therapeutic work with children once I graduate, maybe OT. I want to give children a support system and someone who genuinely wants to help them. I had this growing up with friends, family and a local organization known as the Cody Dieruf Benefit Foundation for CF, and it made the biggest difference in my life. Having this support was incredible for me, and I want to give other children an opportunity to

have that kind of support.

What is life like living on your own? Being independent as a person with CF is hard sometimes. I find it difficult dealing with insurance and doctors appointments and admissions one or two times a year. What I find the most difficult, though, is being out in public – say in a classroom or the movie theater – and always coughing. As a kid you can get away with that more and people may try to not glare or make it noticeable. But as an adult, other people don't mind openly giving you the "stink eye" or commenting, "Maybe you shouldn't be here right now," or "Uhh... can you go wash your hands?!" These types of comments have been made to me and it really is hard to not take it personally and let it affect your self-esteem. I try to shake it off, but some people's comments can be hurtful!

How do you fit in CF meds, studying, classes and sports while in college? Being compliant with medication is very difficult for me, I have a busy schedule! Plus, I am absolutely NOT a morning person. I bet most people with CF can agree with me there! Some days I have to go to class in the morning, then come home mid-day and do a med that was supposed to be in the morning. I understand life happens and if I miss treatments I'm not too hard on myself. All I can do is try my best to get my meds in, because you have to live your life doing fun stuff or else you become miserable and maybe, even, depressed. This is something I went through my second year in college. I focused too much on studying and medicine. And I did not go out and have fun often and I was miserable. Even though I did my medicine more than usual, I became sick more frequently. I believe mental state has a HUGE impact on physical health.

How do you catch up after a hospitalization? Oh man! This hospital admission was my first DURING college classes; usually it's in the summertime. I did some work in the hospital, but that was difficult. I came back and feel that I never really caught up but that I tried my hardest to make up assignments and tests. Fingers crossed I passed the semester! (Kind of joking, I'm a good student so I am sure I passed.)

What motivates you even when feeling sick? I am a stubborn person. I know when I am sick and should take it easy, but I sometimes refuse to let my sick body stop me from pursuing what I love. I push through sports and everyday life until it's time to be in the hospital. It is not the healthiest way to go about it, but to me it is important. I listen to my body more now that I am older, considering I'm not as healthy as I was when I was a young teenager, but I still am stubborn and don't let CF stop me from what I love to do. I just might not push myself as hard.

Is snowboarding a common sport in Montana? I absolutely LOVE snowboarding. I live about 30 minutes from a local ski hill. Skiing and snowboarding are very common here in Bozeman, especially for students. I know some students who come to MSU just for the ski hill! Snowboarding really helps my lungs, even though it is cold. The mountain air is so fresh that it's hard not to enjoy it! It is also a workout! Being up in elevation makes it harder to breathe, but at the end of the day I feel like I just did a breathing treatment. One thing I would recommend, though, when it is cold bring a scarf so you can breathe into that so the icy cold doesn't go straight to your lungs.

How did you get into Roller Derby? I joined the Gallatin Roller Girlz (you can like them on Facebook!) in August. My mother called me up and wanted me to go to a "boot camp" with her. I had no idea what this sport was and I had never roller skated before! But, after a few practices and watching the bouts (games), I fell in love with it. My first bout was May 9! This also helps my lungs quite a bit. The practices work on cardio, so my lungs are working hard (the best they can, I take frequent breaks and use my inhaler often). Also, the sport is very physical! If you don't know what roller derby is, it is physical because we hit each other, on roller skates, HARD. We want the other team to lose, so we knock them down. So, in a sense, after practice it is like I've done a vest treatment!

Tell us about your team. As a Roller Girl, you get to choose specific names and numbers. My name is Drill Bit Messacre. My mother's name is One Hot-Flash Mama. We also get to pick numbers. Since my mother and I are the dynamic mother-daughter derby duo, we chose numbers that correspond with each other. Mine is f508, my mother's is g542x (these are my CF mutations).

What about fundraising? I have participated in two CF walks with a team as well as given speeches at a local foundation that helps the families with medical costs and helps the kids do active stuff if they want. I have given a speech about myself and how I handle CF three years in a row now. I have a little stage fright, but I feel it is important for the community to see the faces of the people they are donating the money to.

In a relationship? Yes, I have been with my boyfriend for almost five years. We met in high school and now pretty much live together. He is very supportive of me and my CF and knows every part about it. He always visits during hospital admissions and is there for venting sessions whenever I need it.

Who makes up your best support? My family is a huge support sys-

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tem for me. My brother has CF as well, so he understands what others cannot. He and I are very close (we actually live together!), and we help each other when needed. I also have a best friend from high school who is an amazing supporter. Her major is microbiology, so she probably knows more about CF than I do (in regards to how the cells are). One thing she is great for is yelling at people who may glare at me for coughing or, if they were to say something, I know she would have my back.

What is the biggest obstacle to making friends in college? In high school, everyone knew about my CF. People talked and I lived in a small town so it was no secret. When I came to college, it was hard for me to tell the people I met. I didn't know when the right time to tell them even was! I have met a few great friends in college, and once I told them about CF they were supportive and understanding. And curious! I like curious instead of assumptions. They now attend fundraisers and visit me when I am admitted for tune-ups.

Any hobbies? I am a pretty active person, so during the summers I hike, go camping, ride my motorcycle (which I have officially name BLAKE), wakeboard, float on the river and play tennis. And of course Derby! Winter is mainly for snowboarding and occasionally snowmobiling. Other hobbies of mine are reading fiction stories, watching movies – I'm a big movie buff – and watching hockey – GO AVS! Of all the sports, hockey is my absolute favorite to watch.

What's your favorite color? Electric blue! No reason, it's just super pretty!

Favorite music? I like all kinds of music, it depends on my mood. But the one band I could listen to any time of day is Mumford and Sons. They are fantastic.

Funniest CF moment? I had one hospital room that someone who painted the wall must have had a sense of humor. The paint looked like a, uhh, "male anatomy" figure. The nurses, RT staff and I had a good laugh about that one! It was kinda hidden, but was obvious what it looked like!

What do you think about the new CF therapies? I hope every day that a cure is found, but I don't think about it often. I try to live in the present because thinking about the future makes me upset.

What advice would you give to someone with CF contemplating college? It is hard, but able to be done. Make sure to have a good relationship with professors so that when you have to miss classes they know why and will be more willing to help. Make friends in classes, too! (Even if they are friends for only that semester.) It is nice having someone from whom you can bum notes for classes that you missed. Most importantly, make sure to have fun! You will have ups and downs, and it is important to keep your mental health stable to keep your physical health stable.

Do you have a philosophy you follow? Live in the now, and do what makes YOU happy, even if others may question it a bit. ▲

Jessika is 21 and has CF. She is a student living in Bozeman, MT. Her contact info is: soccernstage@gmail.com.

Jeanie Hanley is 52 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2. Andrea Eisenman is 50 and has CF. She is a Director of USACFA and is the Executive Editor of CF Roundtable *and Webmaster. Her contact information is on page 2.*

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

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I am courageous.

Overall, I look back at the lives of my CF peers who have pursued a transplant, as well as myself, and I feel grateful to have such personal exposure to heroism. All of us walked to the edge of the unknown and took a leap. We chose to pursue such a risk because we loved life and wanted a second chance. I also have to acknowledge those CF peers who chose not to have a transplant. Accepting this exhausting and exhilarating life with cystic fibrosis as complete is quintessential courage.

In the movie, *The Best Exotic Marigold Hotel*, the main character stated his mantra, "All will be fine in the end. If it's not fine, it's not the end." I like this conviction because it highlights hope, surrender, courage, even maturity. It soothes fear and promotes acceptance. It applies to all of us pre- and post-transplant.

Everyone I've met who is post-transplant has had his or her own path—and own mantras—to survive. If you are living with CF or post-transplant, I encourage you to take pen to paper and write a list of convictions starting with, "I am..." Whatever you come up with will be a glowing reflection of your mighty Spirit. Read and re-read the list, knowing that whatever happens, you can expect these convictions to be your Truth.

Isabel is 43 and has CF. She lives in Redwood City, CA. She works as a bereavement counselor. You may contact her at: Isabear27@hotmail.com

Piper Beatty Welsh

Hello! My name is Piper Beatty Welsh and I am 33 years old with cystic fibrosis and two bilateral lung transplants. I was diagnosed at the age of six weeks in 1981, and I have been a longtime participant in the CF community, particularly through my work with the Cystic Fibrosis Foundation. I grew up in Colorado and enjoyed a relatively healthy childhood (despite the regular in-hospital "tune ups") through high school. As I came into adulthood with CF, especially through college and law school, I realized that I needed to focus on self-education, connecting with the broader CF community, and learning to advocate for myself both as a person and a patient. *CF Roundtable* helped me to accomplish those goals, and I am honored to now serve as a Director of USACFA. I am the grateful recipient of a bilateral lung transplant in 2010 and, after a brief but fierce battle with chronic rejection, a re-transplant on Christmas Day of 2013. I currently live in beautiful Colorado with my husband, Patrick, and our very lively dog, Sampson.

PHOTO BY BRIAN BLOOM PHOTOGRAPHY.

PIPER BEATTY WELSH

I hold a BA from Emory University, a JD from Columbia University Law School, and a Master's in Public Administration (with a focus on nonprofit leadership and management) from New York University's Wagner School of Public Service. In 2011, I was honored to receive the CFF's Alex Deford Award in recognition of service to the community.

Reid D'Amico

Hi all! My name is Reid D'Amico, and I am honored to be a Director of this great organization. I was born outside of Washington, DC, but spent the majority of my childhood in Hilton Head Island, SC. After being diagnosed with CF at the age of 11, I have been fascinated with medical research. I attended Duke University in Durham, NC, and developed this passion by majoring in biomedical engineering. The majority of my undergraduate research focused on stem cells, tissue engineering and regenerative medicine. I graduated from Duke in May of 2015, and in August 2015 I will be starting my Ph.D. in biomedical engineering at Vanderbilt University in Nashville, TN.

REID D'AMICO

I plan to use stem cells and biomaterials to develop the next generation of polymeric biomaterials for regenerative medicine and medical device technologies. My ultimate goal is to apply my findings to pulmonary diseases such as cystic fibrosis. I am excited to expand my involvement with the CF community. I was a recipient of the Cystic Fibrosis Scholarship Foundation and Boomer Esiason scholarships. I also have a column "CF and Regenerative Medicine" with BioNews Services. I am grateful for all of these opportunities, and am looking forward to influencing the CF community by contributing to *CF Roundtable* and working with USACFA.

PROTECTING WHAT MATTERS

Expanding The Discussion Of Transplant

By Mark Manginelli

"Don't take your organs to heaven; Lord knows we need them down here." (If that's too spiritual, use the next quote.) "Organ transplantation is a gift anyone can give. It has no cost and it can be tremendously powerful." According to the CFF.org website and CF Foundation Patient Registry, nearly 2,800 people with CF have received lung transplants since 1990. Being a member of our community provides us a unique perspective of organ donation and transplantation and just how important it is to share their significance both within the community and beyond.

Even though there have been amazing improvements to the care we receive, there may come a time when these therapies offer little to no help and the drugs do only so much; lung transplantation may be the only viable option. Thankfully, we have resources available to get the conversation started. If there becomes even a shadow of a doubt that lung transplantation may be in your future, don't wait. It's so much better to have the conversation with the luxury of time before it becomes too late.

Our amazing CF advocate and friend, Jerry Cahill, shared very insightful words about his transplant experience along with some harsh realities of organ transplantation in general. "Every 80 minutes, somebody on a transplant waiting list dies without getting the opportunity to return to their families, friends and communities and to live a normal, healthy life. Why? Because there is a shortage of available organs."

Jerry firmly believes that spreading the word about organ donation goes hand in hand with raising awareness of cystic fibrosis. People with cystic fibrosis and the members of the CF community truly need to be educated on the progression of CF and the transplant process. Most importantly, our CF centers need to be more proactive in starting the conversation about transplant and everything that comes with it.

Along with the physical and emotional challenges a transplant can bring, many people are often concerned with the dramatic cost of such a procedure. According to a 2014 Milliman research report, the estimated billed charges for a single lung transplant was $785,000 and it rose to $1,037,700 for a bilateral lung transplant. We all obviously hope our health insurances pay for a majority or all of these expenses but, unfortunately, that is not always the case.

Thankfully, there are some amazing resources to supplement these exorbitant costs that are not absorbed by insurance. The first organization that most members of our community have heard of is the Boomer Esiason Foundation (BEF). Jerry is a longtime volunteer of the BEF, and he oversees the transplant grant program. Receiving his bilateral lung transplant on April 18, 2012, he knows first-hand how every new day is a gift. "I think about my donor [and family] every day and I want to make them proud and stay healthy." Last year, the BEF awarded nearly $100,000 in transplant grants.

> *"Along with the physical and emotional challenges a transplant can bring, many people are often concerned with the dramatic cost of such a procedure."*

MARK MANGINELLI

Another organization that Jerry continues to use to this day for his post-transplant costs is HelpHOPELive (www.helphopelive.org). It is a national nonprofit that brings communities together in tax-exempt fundraising efforts to help pay medical bills and access treatments. The main premise behind HelpHOPELive is that it assists you in your own personal fundraising efforts for your transplant fund.

From initial discussions about starting a campaign, through active phases of fundraising and payment of uninsured expenses, the HelpHOPELive team will

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be with you every step of the way. They connect you with one of their expert fundraising coordinators who will work closely with you, your family, and friends to identify a fundraising plan of action that fits each individual's needs. Funds raised through these efforts go to HelpHOPELive where they are held in a regional restricted fund in honor of the patient for the region where the patient lives and are available throughout the patient's life to pay uninsured expenses. The wonderful staff manages all the funds and pays bills directly. Once your fund starts receiving donations, getting expenses paid is as simple as submitting a "Fund Request Form" with the appropriate supporting bill or receipt to HelpHOPELive.

With the assistance of these two funds and starting the conversation with your CF team EARLY, this process won't be as daunting or expensive as one would initially think. Jerry stresses the importance of having these conversations as early as possible. The challenge today is that the conversations are frowned upon until people get really sick and see a transplant team. The problem with that model and utilizing these resources is that there is no longer that luxury of time to raise the necessary funds when the patients are at that stage.

Looking at the bright side, when I asked Jerry how his transplant affected him, he cheerfully replied, "I can breathe now. The process affected me in such a great way; I'm more open with people and so much more giving and accepting. I have a stronger passion for living."

You can learn all about Jerry's story and his incredibly inspiring campaign, "You Cannot Fail: You Are the Hero of Your Own Story," at www.youcannotfail.com. ▲

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

and prolonged life quality in these patients.
http://tinyurl.com/oaekk42

AeroVanc reduces MRSA in patients with cystic fibrosis

The first inhaled antibiotic in development to address the mounting issue of MRSA lung infection in patients with **cystic fibrosis** met the primary endpoint in a Phase II trial, according to a news release from Savara Pharmaceuticals. AeroVanc, the inhaled dry powder form of vancomycin already approved by the FDA to treat MRSA intravenously, reduced MRSA density in sputum and improved pulmonary function, respiratory symptoms and the time to exacerbation and need for other antibiotics. The inhaled antibiotic, delivered in a capsule-based device designed for self-administration, removes the factors of poor lung penetration and systemic toxicities that limit the intravenous drug as a chronic treatment. Cough was the most commonly reported adverse event; no difference was seen between dose levels or between therapy and placebo groups. Symptoms consistent with bronchoconstriction were observed more frequently in patients treated with AeroVanc 64 mg. All MRSA strains were susceptible to vancomycin.
http://tinyurl.com/nmceby3

PARI Pharma's Vantobra Granted Marketing Authorization in Europe

PARI Pharma's Vantobra®, a new highly concentrated tobramycin nebulizer solution for inhalation delivered by a Tolero® nebulizer, was granted Marketing Authorization from the European Commission. Vantobra is a breakthrough for cystic fibrosis patients as it offers reduced treatment times of four minutes with treatment effects comparable to TOBI®. "In clinical trials, patients were able to take a full treatment of Vantobra within four minutes. Vantobra is a proprietary aqueous solution of 170 mg tobramycin/1.7 ml for inhalation use via an optimized, drug-specific Tolero nebulizer handset. Vantobra and the Tolero handset are packaged together for patient convenience.
http://tinyurl.com/ow3579e

Researchers Use New Method to Predict Pancreatic Damage in CF Patients

In a new study entitled "**Ultrasound echo-intensity predicts severe pancreatic affection in cystic fibrosis patients**," the authors suggest ultrasonography is a suitable and effective diagnostic tool to evaluate pancreatic function in cystic fibrosis patients. The findings could help physicians better diagnose and treat the pancreatic issues and conditions in CF patients. Trans-abdominal ultrasound is a radiation-free technique that allows repeated examinations. Additionally, it's a low-cost exam and the low impact on patients' wellbeing makes it one of the most suitable diagnostic tools for routine follow-up on CF patients.
http://tinyurl.com/oacha9c

Microbial Changes in Patients with Cystic Fibrosis Associated with Pulmonary Exacerbations

Changes in microbial community structure in patients with **cystic fibrosis** are associated with some but not all pulmonary exacerbations. Daily airway bacterial community structures in CF are relatively stable during periods of clinical stability. However, the onset of symptoms of exacerbation may be heralded by marked shifts in these communities, even in the absence of viral infection or antibiotic therapy for the treatment of exacerbation. Monitoring of airway microbial community structures may identify

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PARENTING

God Bless The Broken Road That Led Me To Motherhood

By Jamie H.

About Me

I was diagnosed with cystic fibrosis when I was six months old. I have the delta F508 mutation. Despite following every direction of my doctors throughout my life, in 2010 my lungs began to fail rapidly. My last day of full-time employment was on September 29, 2010. In mid-December, unable to breathe on my own, I was put on a ventilator at the University of Minnesota Medical Center. I have experienced many hospitalizations throughout my life, but nothing is more life changing than not being able to breathe—to be supported in life by a ventilator.

On December 21, 2010, the ninth day of ventilation and only hours before my doctors would have to decide that we would need to end the support, my life-saving lungs came to me. They came from Kansas, where a 17-year-old boy named Landon was in a terrible car accident. His tires caught the soft sand of a dirt road and pulled his truck into the ditch, where it rolled. He was ejected and hit a tree. His family, who is now also my family, made the decision for him to be an organ donor after he was pronounced brain dead. His lungs were healthy and a match for me. My life was forever changed. My life was saved and I have been breathing easy ever since.

The Desire to Be a Mom

Ever since I can remember, I have wanted to be a mother. Many tears have been cried over the desire to be a mother. A lifelong dream was shattered by a terrible disease that would never allow my body to become pregnant. Lung functions for me were below 50 percent for much of my life. I was on more rounds of powerful antibiotics than I can recall. I never weighed more than 100 pounds, as every ounce of energy was burned up breathing and coughing. My body was never fit enough to carry a child let alone have the stamina to take care of all the needs of a child. I grieved this dream and buried it. Each baby I saw out in public would always send a stabbing pain to my heart. The longing for a child really never leaves. I hated knowing that I would be an amazing mother, but I knew this would never be for me. I married a man with a child. I always felt good about that, as I would never take away his dream. He already had a child. I was a stepmother.

The Journey

After the transplant, these dreams resurfaced. I was healthy, breathing and thriving. Those tears returned and once again I desired to be a mom. I have three sisters, two of whom offered to be a surrogate mother for us. We began exploring this option and made an appointment with a fertility specialist in my area. We were excited and nervous about finding out the steps for this process.

JAMIE H. 28 WEEKS PREGNANT.

This appointment that I was looking forward to was shattered quickly. The doctor we spoke with began crushing our dreams of creating a child of our own. My husband is a wonderful father to his daughter. I always knew whatever were to happen, our child would be loved and cared for. We each have a supportive family that would always be there for him or her. I have every intention of fighting as hard as I can to be there as long as I can.

We had many painful discussions about my future. I know this transplant will not last forever. My sisters were more than happy to sacrifice their bodies to assist with this dream. The doctor began to tell us that he would not help us. Our child could not only have CF, he said but also stated, and I will never forget these words, "I am in the business of making families, and with your uncertain future I cannot and will not help you."

I remember staring blankly at the wall, tears streaming down my face. My husband at least had the right mind to say, "Who's to say anyone's life is guaranteed?" The doctor only responded with, "That is unpredictable, your future is predictable." We left, broken hearted.

After the initial shock ended, we eventually made an appointment with another fertility specialist at the University of Minnesota. He did not

have any judgmental views and very professionally told us the steps to make this happen. But it would cost many thousands of dollars: around $30,000. That kind of money was maybe doable with loans, but what about the unpredictable outcome of how the pregnancy would go and the health of one of my sisters? Surrogacy was not covered by my sister's insurance or ours. We wanted to make this happen but needed to figure out all the legal details that went with it.

In the meantime, my husband and I attended adoption classes with a local adoption agency. Again this process is very overwhelming, since it could take years to adopt a child and, again, a lot of money.

A short time later, my grandmother died. She was a hardworking farmer's wife who took care of 17 foster children in addition to the five children of her own. We thought, maybe we should foster. We attended a foster-to-adopt information class with our local county. My extended family knew we had this desire.

Out of the blue one day, a social worker from the county called me and said that my cousin had stated that we would take in children who were in the family and needed a foster home. To make a long story short, we said, "Yes."

We began the process to quickly get approved to take these children into our home. We were ordered to get car seats and a crib and to take pictures of the room, as the date was nearing that the court was to decide the fate of these children. With the support of my family, we pulled it off. We had a nursery all ready for twin girls. Items were donated from friends in just a few short days. We were approved and just waiting for the judge.

A day before the court date, the twin's maternal grandmother came back into the country. The judge told her she would have no rights to the girls if she did not take custody. She was not ready to do this but said, "Yes," just in fear of not seeing them again. English was not her first language, and I still think she did not understand everything at the time. The judge in the last hour ruled for her to take custody as she was a blood relation. She had no room for them, did not speak English, worked full-time and did not have a good relationship with her daughter but was given the blessing to raise these one-year-old twin girls. We were not blood relatives, as the twins were the children of a step-son of someone in our family. Just like that, the dream was taken away. Again we grieved. I cried. I was mad at our foster system. This anger pushed me forward.

The Decision

If a woman addicted to drugs and alcohol was allowed to have children, why wasn't I? I began to do research. Through Facebook groups, I met several women who did just that – have a child after a lung transplant. I was always told this was not possible as the medication we take for rejection was harmful to a child in utero. I never wanted to risk it or risk that a pregnancy would cause me to reject these new lungs. But now I was communicating with women who had had healthy pregnancies with the outcome of healthy children.

I also have met several women who have lived 20-plus years with their transplanted lungs. My biggest fear in life was to be one of those women. I did not want to be here 20 years from now, childless. I wanted to know more, so I contacted the National Transplantation Pregnancy Registry (http://www.ntpr.giftoflifeinstitute.org/). They answered many of my questions, and I learned that I am already on the combo of medications that would be safe for a pregnancy.

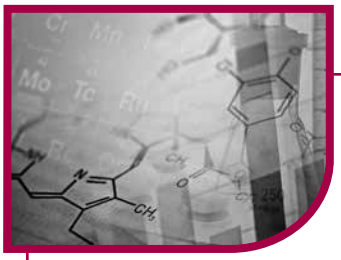
Celcept, one of the popular rejection meds indeed causes miscarriages. But I already was taken off of that and put on Imuran and Prograf. These meds are taken by many other organ recipients who went on to have successful pregnancies. I then took all my new information to my pulmonologist. Although I was always told pregnancy was not an option, I now challenged my doctor with the question, "Why?" and "How can we make this happen?" He knew I was serious and started to make a plan for tests that needed to be completed to prove that I was healthy enough. My heart checked out, my kidneys checked out, and my CMV (cytomegalovirus), which I had contracted from my donor, was under control. I was now making antibodies to fight against it, and it would no longer be harmful to a child that I was carrying. My diabetes and blood pressure were under control. I kept passing the tests!

Next Steps

Since I have CF and am 34 years old, I knew I would have to act fast as time was running out. My husband and I made an appointment with the fertility specialists again. I had very irregular periods post-transplant, and I knew the thick cervical mucus would work as a barrier for sperm to pass through my cervix and through the tubes to be able to get pregnant. Thankfully my specialist agreed to help us as long as I had the support from the Fetal Maternal Medicine team. I was able to get an appointment with them quickly.

We were told of all the risks involved in me carrying a child. They said there was a 50 percent chance of death. Did this freak me out? Yes, of course! Did I think this was the best percentage they could give me because they are not fortune-tellers and 50 percent covers the risk? Yes. I knew I would be a high-risk but I knew in my heart I could do this. I prayed to God and asked that he make this happen

Continued on page 34



SEARCHING FOR THE CURE

Update On Orkambi Clinical Trial

By Meranda Sue Honaker

Vertex's combination therapy, Vx-809/Vx-770 (lumacaftor with ivacaftor) now known as Orkambi, which targets CF patients who are homozygous for the F508del mutation, is pending FDA approval. The Phase III clinical trial of Orkambi met primary endpoints with statistically significant improvements in lung function (FEV_1) in people with CF who have two copies (homozygous) of the F508del gene mutation.

According to Vertex, there were over 1,100 CF patients who participated in the Phase III TRAFFIC and TRANSPORT studies. In the 24-week treatment arm, patients had a mean absolute improvement of FEV_1 with a range of 2.6 to 4.0 percentage points with a mean relative improvement of 4.3 percent to 6.7 percent. Additionally, the pooled analysis of the Phase III studies showed a pulmonary exacerbation reduction rate of 30 and 39 percent for those on combination therapy versus placebo. Finally, of the 1,100-plus patients participating in the study, there was a 4.2 percent rate of discontinuation for patients receiving Orkambi due to adverse events versus 1.6 percent of patients receiving placebo.

On May 12, 2015, Vertex met with the FDA's advisory committee to discuss the new drug application (NDA) for Orkambi in people with CF who have two copies of F508del. This coincided with a public hearing before the advisory committee wherein CF patients, medical providers and advocates were selected by the FDA to discuss their experiences with Orkambi and to advocate for FDA approval. I personally spoke in favor of FDA approval of Orkambi as a study participant who has benefited from this therapy. Speaking to the FDA on behalf of Orkambi approval was made with great consideration. I had only four minutes to convey 18 months' worth of personal experience on Orkambi to the advisory committee. Moreover, the hearing was in a crowded room, and many others were watching via a live stream on the Internet. After hearing from multiple speakers and after reviewing the Orkambi Phase III clinical trial data, the FDA advisory committee voted 12-1 in favor of recommending Orkambi for approval. Currently, the FDA has until July 5 to make a final decision on the approval for marketing Orkambi in the United States for those who have two copies of F508del. Perhaps by the time this article is published in *CF Roundtable,* many of you will have already received your first dose of Orkambi! *[Editor's note: Orkambi received FDA approval on July 2, 2015.]*

Thanks to Orkambi, I have experienced a reduction in exacerbations and fewer episodes of hemoptysis, and I have been able to delay a fourth sinus surgery an additional two years. The first six months of this study were "double blind," meaning my study doctor (Principal Investigator) and I did not know if I was receiving placebo or active study drug. After the first six months ended, I enrolled in the optional "open-label" extension study to monitor long-term safety and efficacy. I remain blind as to the dosage of Orkambi I am on, but I know I have been on Orkambi for over a year through the open-label extension study.

> *"It is up to us as patients to participate in studies that enhance scientific knowledge leading to therapeutic advancements."*

MERANDA HONAKER

I suspect I was not on placebo during the six month blinded phase of my study. Prior to joining the Vx809/Vx770 trial, I had been suffering chronic daily migraines for several weeks, which I experience only when in need of sinus surgery. However, within the first week of beginning Orkambi (or placebo) my sinus migraine completely disappeared. I had not changed anything else in my medical regimen. For the first time in several weeks I was able to open the blinds to allow sunshine in and did not need

to lie down for hours with freezer packs over my face to combat sinus pain. Moreover, my lung congestion improved. Specifically, my sputum production reduced and thus my cough also lessened. Furthermore, I have had one pulmonary exacerbation requiring intravenous antibiotics since joining the Orkambi study. I remain on all of my CF therapies (including a daily IV antifungal) that I was taking prior to joining the Orkambi clinical trial. Orkambi has improved my quality of life and is a great addition to my daily medical regimen.

There are numerous CF studies currently enrolling CF patients in clinical trials. It is up to us as patients to participate in studies that enhance scientific knowledge leading to therapeutic advancements. I encourage all with CF to participate in clinical trials because it benefits the entire community and the future of our healthcare.

Clinical Trials Currently Enrolling:

Visit www.cfroundtable.com/announcements/clinical-trials/ for a comprehensive list of currently enrolling CF studies.

Parion Sciences: Clearing Lungs with ENAC Inhibition in CF (CLEAN-CF): https://www.clinicaltrials.gov/ct2/show/NCT02343445?term=NCT02343445&rank=1

Effect of Chronic Incretin-Based Therapy in CF (CF-Related Diabetes Study) https://www.clinicaltrials.gov/ct2/show/NCT01879228?term=nct01879228&rank=1

Blood Flow and Vascular Function in CF (Exercise, Blood Flow & Artery Function): https://www.clinicaltrials.gov/ct2/show/NCT02057458?term=nct02057458&rank=1

Comparison of Absorption of Vitamin D (Powdered Pill Vit D versus Oil Form): https://www.clinicaltrials.gov/ct2/show/NCT01880346?term=nct01880346&rank=1

Gender Disparity and Hormones in CF (Investigating Impact of Hormones in CF): https://clinicaltrials.gov/shows/NCT02036879 ▲

Meranda is 32 and has CF. She is a Director of USACFA and is the Vice President. If you are a CF clinic and would like to contact her to list a CF clinical trial on the CF Roundtable *website, e-mail her at: MHonaker@usacfa.org*

Benefactors

BRONZE

Susie Baldwin (In honor of her lung donor)
Irene Bernaix (In honor of 50th anniversary of Paul and Kathy Russell)
Robert and Nancy Coleman
Edward & Elaine Corr
Pauline DiNello
James and Ann Nash
Sidney Rabinowitz (In memory of Ilana Davida Schwartz)
Dina Shingleton
Ronni Wetmore

PLATINUM

Abbvie
Boomer Esiason Foundation
Cystic Fibrosis Foundation
Estate of Pamela Gordon Beaton

Cystic Fibrosis Mothers

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

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

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

WELLNESS
N OF WHAT?

By Julie Desch

When President Barack Obama announced his $2.15 million precision medicine initiative during the State of the Union address this year, the CF community was elated. The words "cystic fibrosis" were spoken by our President as an example of how medical care and research are changing, and he used the Kalydeco experience as an example. As we all know, a small percentage of patients with the G551D gating mutation benefitted tremendously from Kalydeco, while those with other classes of mutations didn't. So the FDA approved the drug for this subset of patients, rather than all CF patients in general (since then, other gating mutations have been added to the list). President Obama used this example to illustrate how we need to learn more by extensive genetic and other data collection in order to fine tune our medical prescribing and approval system in order to improve our national healthcare.

Let's think for a minute about how classical clinical research is done traditionally. Let's say Drug A has been developed and we want to see if it is effective in Disease B. In this case, after proving safety and finding the appropriate dose of Drug A, thousands of people with Disease B will engage in Phase III clinical trials for a significant period of time, and data is collected. If Drug B is found to be "effective enough," based on predetermined endpoints, the drug is approved. But there are likely huge swaths of people for whom drug B did nothing and, perhaps, even a few where it caused serious problems. At the other end of the bell curve lie the people for whom Drug B was phenomenally effective... way higher than the average. It's basically a statistical game, and when enough people fall beneath the bell curve of "effectiveness," the drug is approved.

But what about the outliers—the people at the end of the bell curve, where the drug did nothing or, in fact, the drug was harmful? In these folks, there were most likely genetic, epigenetic or environmental factors that were not seen (because they were not looked for), and with this experiment design, we learn nothing about why the drug didn't work for them. Nor do we learn anything about the "super-responders" at the other end of the curve. What was it about them that made the drug so effective? Perhaps if we understood this, even more effective drugs could be developed.

There is a type of study design that gets at these issues and is becoming of more interest as scientists are understanding that "treatment by the average effect" is less than ideal. The study design is termed "N of 1," which is basically an experiment where an intervention is given to one person, who serves as his or her own baseline control. If you think about it, we in CF-land do this all the time. We decide, hopefully with our doctor's approval, to do a trial of an inhaled medication and see if it works for us. For example, Pulmozyme is effective for me, but for some of my friends it causes hemoptysis. So I decide to use it, and my friend doesn't. Pulmozyme is approved for everyone with CF, but there are a few people who can't use it. How did we learn this? N of 1 trials. But this example is more of a random event, where a patient just discovers something informally, i.e., "This drug makes me bleed!"

JULIE DESCH, MD

What if we were to design N of 1 trials with the advent of CFTR modulator drugs approved (Kalydeco) and in development (Orkambi and others)? Early in clinical trials, Kalydeco was used on delta F508 homozygotes like me. In fact, I was in such a trial. At the end of the study, the averages showed that Kalydeco didn't work. It didn't meet its endpoints. Yet, it is known that a small population of people who are double delta F508 have "residual function." For some reason, they actually do have a small amount of functional CFTR at the apical membrane, and for these people, Kalydeco may in fact work. But these people are ignored in the standard study design, because they were at the extreme end of the bell curve that (statistically) showed no effects. An N of 1 study design would identify these people.

Even with inhalation of antibiotics other than TOBI, Cayston or Colistin (FDA-approved antibiotics for inhalation), N of 1 studies could identify sig-

nificant benefits that would improve the lives of some. Heck, we all inhaled tobramycin before the advent of TOBI, right? These were (informal) N of 1 experiments that were done which then led to the development of TOBI! I have friends whose docs prescribe inhaled (but "off label" because these are meds designed for IV use) cephalosporins, or other classes of antibiotics, and they are helpful! My physician won't consider it because they are not approved and, therefore, not covered. And why are they not approved? Because there would need to be large, Phase III randomized, placebo-controlled studies done that showed efficacy and safety. If the multiple people who have success with inhaling meds that are not yet approved for inhalation had a formalized way of showing their effectiveness (pooled N of 1 trials), I would have the ammunition to go to my doctor and say, "Let's try this!"

N of 1 trials are making a huge impact in a community known as Quantified Self. Their tagline says it all, "self-knowledge through numbers." This is a large and growing group of passionate (and somewhat nerdy) people who love to use wearable devices and other new technology to measure aspects of their own bodies and lives in order to gather data that is useful to them. Through this data, they learn about what does and doesn't affect their health, for instance, by creating their own N of 1 experiments. They then present their findings at meet-ups and even an annual national convention. If you want to be fascinated for hours, check out the Quantified Self website, where you can lose yourself watching video presentations showing how people learned to improve their lives and health through self-measurement. Or...maybe that's just me.

N of 1 trials are coming to you soon! They have to. The new drugs in development are going to be so finely tuned that these types of trials will need to be designed in order to prove efficacy. If you have an extremely rare mutation, large Phase III clinical trials are impossible...there just aren't enough people. My advice is to start playing around with your own N of 1 experiments. Start collecting data (my favorites are resting heart rate and mood scale, but heck, go all out and add a few more things) and then when you have a stable baseline, add an intervention... for example, a daily walk or meditation. See what happens to your baseline numbers. Become your own best scientist! ▲

Julie is 54 and is a physician who has CF. You may contact her at: jdesch@usacfa.org.

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signatures that could be useful in predicting CF respiratory exacerbations, which in turn may enable more prompt and/or appropriate therapy for exacerbations.
http://tinyurl.com/nhnmu83

Clinical Study Details Pulmonary Function Improvement In Five CF Patients Using AffloVest

AffloVest, a wearable, self-contained oscillation vest device from International Biophysics that helps patients with cystic fibrosis breathe easier, is now proven in a new clinical study that it increased forced expiratory volume in one second (FEV_1) an average of 11.5%.
http://tinyurl.com/ox629wl

Regenerative Medicine, Stem Cells Poised to Impact Development of CF Treatment

Regenerative medicine is the practice of delivering cells and cell products to diseased or damaged tissue. In diseases like cystic fibrosis, the lungs and pancreas accumulate damage due to thick mucus secretions and chronic infections. The current treatment of antibiotics, airway clearance, mucus alteration, CFTR modulation and pancreatic enzymes only help to manage symptoms. Even with the current work to correct the defective CFTR protein that leads to the diagnosis of cystic fibrosis, there is insufficient evidence to conclude if new medications will halt the progression of the disease. However, regenerative medicine offers the possibility of controlling the development of this damage.
http://tinyurl.com/qa3dllx

Advanced Inhalation Therapies Granted European Orphan Drug Designation for Nitric Oxide for the Treatment of Cystic Fibrosis

Advanced Inhalation Therapies Ltd. (AIT) announced that the European Commission granted orphan medicinal product designation to AIT-CF, the company's proprietary high-dose formulation of nitric oxide (NO), for the treatment of cystic fibrosis. As a naturally occurring chemical in the body, NO, when inhaled at high concentrations, kills bacteria and fungi in the lungs of CF patients and has shown broad anti-infective activity.
http://tinyurl.com/p3x8t9k

Vertex Announces Data Presentations at European Cystic Fibrosis Society (ECFS) Conference

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only if it was meant to be. We decided to proceed.

My fertility specialist put us on the fast track. We started fertility medication to help me ovulate. We agreed that there were to be only one mature follicle as a result of the medication to decrease the chances of me becoming pregnant with multiples. On the fifth round of fertility meds and one week before I turned 35 (I told myself I would try only up until my 35th birthday), I got a positive on a pregnancy test.

Once I saw that positive, a wave of worry came over me. I was consumed with anxiety of whether or not I had made the right decision. Would I really be okay? Would my child be okay? I was frantic, to be honest. On a Facebook support group, a woman shared this quote with me: "Don't let the fear of what if, rob you of the joy of what is." It calmed me. I wrote the quote down in big letters and taped it to my refrigerator. It was my mantra for the coming months.

The Pregnancy

My pregnancy was pretty uneventful (except that I had a doctor's appointment every week for one specialist or the other). We were keeping a close eye on my kidneys and my blood pressure. I have had high blood pressure since transplant, and I have also had CF-related diabetes since I was 16. (Another risk factor.) My diabetes management had never been better, with an A1C of 5.5. My kidney levels were elevated a bit but never at a dangerous level. But my blood pressure was getting harder to control as the medications I used before pregnancy to control my blood pressure were not safe to use during pregnancy and the current medications were not controlling it as well. I was able to maintain my blood pressure under the recommended range for pregnancy. My lung function continued to remain stable at 102 percent.

I had a heart-to-heart with my doctors. I wanted to make sure that they knew that I am forever grateful for their help in saving my life. They got me on that transplant list and made the transplant happen. I wanted them to know I did not take that for granted and did not want to be known as noncompliant and, if for some reason, some day I would need another transplant, I didn't want this to be a factor. Thankfully my doctors were understanding and told me that I have always done everything they have asked of me. I did my research and made an informed decision. One doctor told me that we do transplants so you can live your life the way you always wanted. My team of specialists kept a close eye on me throughout my pregnancy. The baby looked great, was healthy and was growing as she should. But no one predicted that my placenta would tear in the middle of the night. At 28.5 weeks, my placenta tore and my husband rushed me to the hospital. I also started contractions. I was losing blood fast and there was no choice but to deliver my baby via C-section.

> *"Ever since I can remember, I have wanted to be a mother."*

Elaine

She was born at 2 pounds 11 ounces. She is thriving. She is my joy. She is my love and reason to live. My husband is so very supportive and also completely madly in love with our sweet baby. I have been able to nourish her with my breast milk. She is perfect in every way. She is just little but will be coming home with us very soon. I have recovered from the traumatic birth. I did spend nine days in the hospital after my C-section. I developed preeclampsia after delivery and developed a mild infection at one of my IV sites. Since then my labs have returned to baseline.

I lost my baby weight quickly and that is my biggest challenge – keeping on the weight while breastfeeding. But I am realistic and made the compromise that, if my weight falls below 100 pounds, I will stop. My husband and I are looking forward to the day that we can take her home, which should be in the next week or so! I will never be ashamed that I went for it. Others may never understand why I was willing to take these risks. I made my dreams come true. I am confident still that she will be well taken care of no matter what. The same what ifs do constantly play in my mind, and we will cross that bridge together, if and when it comes, as a family. We will do it together.

I am proud to say I am the 28th woman on record of having a child in the U.S. and Canada after bilateral lung transplant. I wouldn't change a thing. My donor family is just as excited. My donor's mom calls herself Nana and his father calls himself Papa. My donor's uncle wants to be called Uncle Bubba. They are all planning a trip to Minnesota very soon to meet the newest member of the family. As a side note, Elaine was born on National Donate Life day, 2015. My lung donor's sister had a baby two years ago on National Donate Life day as well! We don't think it was just a coincidence ! We have someone who is always with us letting us know.

After all, this little miracle that we named Elaine would not have been possible without the gift of lungs from Landon. I have faith that this little miracle was meant to be and I will be able to enjoy many years with her making memories. ▲

Jamie is 35 and has CF. She and her husband, Chris, live in Minneapolis, MN. You may contact her by e-mail at: Ruby101679@aol.com or by phone at: 612-670-1710.

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Vertex Pharmaceuticals Incorporated announced data from PROGRESS, the long-term extension study of the investigational regimen ORKAMBI™ (lumacaftor/ivacaftor). One thousand, thirty-one people who completed 24 weeks of treatment in either of the Phase III studies, TRAFFIC or TRANSPORT, entered the 96-week PROGRESS Phase III extension study in which everyone received one of two lumacaftor/ivacaftor combination regimens. The data showed that the initial improvements in lung function (percent predicted forced expiratory volume in one second, or $ppFEV_1$) observed in the 24-week TRAFFIC and TRANSPORT studies among those treated with a lumacaftor/ivacaftor combination were sustained through 48 weeks of treatment across all patients. Reduced rates of pulmonary exacerbations and improvements in body mass index (BMI) and patient-reported respiratory symptoms as measured by the respiratory domain of the Cystic Fibrosis Questionnaire Revised (CFQ-R) were also maintained over 48 weeks. At the time of this analysis, no new safety concerns were identified. Over 48 weeks, the most common adverse events were infective pulmonary exacerbation, cough and increased sputum.
http://tinyurl.com/pgh745k

Tiny Corbus looks to take on Vertex with experimental lung drug

Corbus Pharmaceuticals received $5 million from the Cystic Fibrosis Foundation last month to develop the drug Resunab, and expects to begin a mid-stage study this quarter. Resunab aims to manage the inflammation the disease causes.
http://tinyurl.com/p3elrdn

Corbus Pharmaceuticals Announces Presentation of Positive Data on Resunab™ in Pre-Clinical Models of Cystic Fibrosis at the 2015 Cystic Fibrosis Foundation Research Conference

"Resunab Benefits in The Murine Model of CF Lung Infection and Inflammation without Jeopardizing Resolution of Pseudomonas Aeruginosa (PA) Colonization in the Lung" indicates that in CFTR-deficient mice infected with Pseudomonas aeruginosa, Resunab improved survival, decreased weight loss, reduced the numbers of neutrophils and white blood cells in the lung and improved the ability of animals to resolve pulmonary infection as assessed by lung bacterial colony forming units ("CFUs"), compared to control treatment. The study suggests that Resunab could potentially be effective

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MILESTONES

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

ANNIVERSARIES

Birthday

LoriAnn Reynolds, 42
Las Vegas, NV
May 15, 2015

Kathy Russell, 71
Gresham, OR
April 17, 2015

Michael Schnitzer, 58
Vernon Hills, IL
June 4, 2015

Wedding

Kathy & Paul Russell
Gresham, OR
50 years on March 27, 2015

Michael & Shari Schnitzer
Vernon Hills, IL
30 years on April 28, 2015

Transplant

Andrea Eisenman, 50
New York, NY
Bilateral lungs
15 years on April 25, 2015

Jess Newport, 28
Durham, NC
Bilateral lungs
5 years on April 18, 2015

NEW BEGINNINGS

Transplant

Michael Schnitzer, 57
Vernon Hills, IL
Bilateral lungs
May 28, 2015

CONVERSATION CORNER

Sleep Study

By Andrea Eisenman

I come from a long line of loud snorers. It probably has something to do with our big noses and/or small chins. After my initial meeting with the sleep study doctor at NY-Presbyterian Medical Center, I learned that a small chin may cause the mouth to drop open and cause snoring. The snoring in itself is not the main problem, unless you keep up half the neighborhood. It is the sleep apnea that can accompany the snoring, or rather, when the snoring stops and one has stopped breathing temporarily through the night of sleep.

Sleep apnea can cause minor problems such as headaches and feeling tired all day to heart attacks and strokes. I know I had sleep apnea years ago pre-transplant when I would wake up with a headache or woke up to take in a big breath, feeling like I had not been breathing for some time. I know I had oxygen deprivation, but was it from my apnea or just my debilitated lungs? I was constantly tired and felt bad, but I was also pretty sick while waiting for a transplant for a couple of years.

Recently, about six months ago, my husband told me that I snore. Loudly! He prods me and asks me to roll onto my side where I usually will stop snoring. But I find that sleeping on my back is the most comfortable. So, even when I roll onto my side, I am lying on my back in no time and snoring once again.

One day he decided to record me with his cell phone while I was snoring, to prove to me how loud it was and that he felt I had sleep apnea. After laughing hysterically at my own snoring, it was obvious that I stopped breathing a few times. I realized that I better mention it to my transplant pulmonologist at my clinic. I knew what was coming, I would need to do a sleep study and might have to use a CPAP (continuous positive airway pressure) machine. This keeps the airway from closing when the tissues start to collapse around one's throat.

The sleep study is used to determine how severe the apnea is and whether any medical interference is necessary. Usually, when a person lies on his or her back, the muscles in the throat relax too much and cause closing of the larynx and soft tissue that block the passage of air. When this occurs, people can stop breathing temporarily. Luckily, there are a few options to help fix this. The Positive Airway Pressure (PAP) devices are used for those with moderate to severe apnea and are the most popular option. This machine pushes air through one's nose that then keeps the throat from closing. There are also dental plates that can be adjusted by a dentist, plus some home remedies for those with minor issues.

It turns out, I have sleep apnea only when on my back, in REM cycle sleep. Easy. Simple. Just sleep on my side or stomach, right? No. I try to but always end up on my back in the morning, mouth agape.

Some home remedies include pushing one's bed next to a wall, so that a person sleeps with his or her back up to the wall. Not sure what keeps a person from turning onto the back. Then there is the wearing of a knapsack filled with a few tennis balls. This prevents people from lying on their backs because who wants to sleep on top of tennis balls protruding into their spine? But then again, who can sleep while wearing a knapsack in bed?! Finally, a body pillow was suggested to keep a sleeper on his or her side or at least make it more comfortable. This is the option I am pursuing. Due to my back problems—spinal fusion, hip bursitis and stiffness from spine surgery— clutching a soft body pillow might help me stay on my side and alleviate the pain I sometimes experience from sleeping on my side.

Because I am also having sinus surgery soon, we will re-evaluate with a take-home pulse-ox device after surgery to see if I have mastered sleeping on my side, and whether I still have loss of oxygen due to apnea while sleeping. Once that is determined, I may have to use a CPAP device after all. But in the recent months, I walk around tired most of the day and my memory is extremely sketchy. The doctor told me it was because I am not breathing properly when sleeping on my back, which currently is most of the night.

While I do not welcome sleeping with a machine blowing air up my nose, or any other body part, if I can have a day where I do not feel like I need a nap by 1 p.m., I would welcome that apparatus with open nostrils. ▲

Andrea is 50 and has CF. She is a Director of USACFA and is the Executive Editor of CF Roundtable *and Webmaster. Her contact information is on page 2.*

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in addressing inflammation in cystic fibrosis patients, and furthermore improve the body's ability to resolve pulmonary bacterial infection.
http://tinyurl.com/o7d9t48

Parion Sciences Announces Initiation of Phase II Clinical Study of P-1037 for the Treatment of Cystic Fibrosis

Parion Sciences announced that it has begun enrollment of a Phase II clinical trial of P-1037 in patients with cystic fibrosis (CF). The trial has been named the "CLEAN-CF" trial which refers to "Clearing Lungs with ENaC inhibition in Cystic Fibrosis." The CLEAN-CF study will include CF patients regardless of an individual's genetic mutation. Inhibiting the epithelial sodium channels (ENaC) in the airways with P-1037, an "ENaC blocker," is expected to re-hydrate the mucus layers, thus restoring airway clearance, improve lung function and, ultimately, reduce exacerbations. P-1037 has demonstrated to be long acting in preclinical models and to have a favorable safety and tolerability profile in the completed Phase I studies. Parion expects to enroll approximately 120 patients at 30 sites.
http://tinyurl.com/ovrfhok

New drug "could ease symptoms of cystic fibrosis"

Scottish researchers believe a new drug could ease the symptoms of people with cystic fibrosis. Lynovex, developed by Aberdeen-based bio-technology company Novabiotics, is one of a number of new drugs being worked on to tackle cystic fibrosis. Lynovex controls the bacteria and fungi that causes lung infections and could help the lungs to work effectively for as long as possible as it breaks down the sticky mucus in the lungs of patients. The active component of the new medicine is Cysteamine, a drug used for 20 years to treat an unrelated rare condition called cystinosis. The fact that it is already approved for clinical use means it could be quickly approved for use on people with cystic fibrosis.
http://tinyurl.com/pnw4a2u

Protalix BioTherapeutics Announces AIR DNase™ Data Presented at the 38th European Cystic Fibrosis Conference

AIR DNase is the company's inhaled, chemically-modified, plant cell-expressed recombinant form of human deoxyribonuclease I, or DNase, that is resistant to actin, a potent inhibitor of DNase that is found in high concentrations in the lungs of CF patients. AIR DNase works by cleaving extracellular DNA and thinning the thick mucus that accumulates in CF patients' lungs.
http://tinyurl.com/pwu3srd

Female cystic fibrosis patients need more contraceptive guidance, study finds

Only half of women with CF report using contraception and frequently are apt to become pregnant unintentionally. Researchers used a survey to assess **reproductive health** and quality of life in patients with cystic fibrosis. Female participants aged 18 to 45 years were recruited. Among 53 women surveyed, 83 percent reported being sexually active and 27 percent said they had been pregnant. Twenty-two percent of reported pregnancies were terminated, due to either unplanned pregnancy or suboptimal health status. Forty-nine percent of participants said they use contraception, compared to 65 percent of women in the same general U.S. population. Furthermore, women with more severe CF disease were revealed to be less likely to use contraception. **Condoms** and **oral contraceptive pills** were the most commonly used methods.
http://tinyurl.com/q7fvx87

Common Bacteria That Causes Pneumonia Found to Be Rare in CF Patients

Researchers have recently published findings indicating that lung colonization by the bacteria *Streptococcus pneumonia* is relatively rare in patients with cystic fibrosis (CF) and, when present, it apparently does not have an adverse impact on patients' clinical conditions. The study is entitled "**Prevalence and impact of Streptococcus pneumonia in adult cystic fibrosis patients: a retrospective chart review and capsular serotyping study**." The research team concluded that in the Canadian adult CF patient cohort analyzed, incident Pneumococcus infection is relatively rare and it is not linked to clinical deterioration of the patient nor to chronic infection.
http://tinyurl.com/ouxo664

Cystic fibrosis-related diabetes requires different approach

Cystic fibrosis-related diabetes (CFRD) is a unique disease, and it requires a different mindset on the part of the treating physician. Patients with CFRD do not appear to get macrovascular complications. Diabetes is the most common comorbidity associated with CF. It's associated with pancreatic insufficiency, liver dysfunction, requirement for corticosteroids, and prognostically with undernutrition, worse pulmonary function and early death. The prevalence of CFRD is age-related. It's rare in children, but the prevalence climbs to about 15% in adolescents, 40% in 20- to 39-year-olds and 55% after age 40. The definitive treatment for CFRD is insulin. Multiple studies have demonstrated that diabetes has a negative impact upon survival in patients with CF. Both hyperglycemia and insulin insufficiency have negative impacts on the CF lung disease. Aggressive screening and early initiation of insulin therapy help reverse chronic weight loss and reduce mortality in patients with CFRD.
http://tinyurl.com/oywp5lz

Celtaxsys Inc. Receives $5M Research Award to Advance Once-Daily, Oral Anti-Inflammatory Medicine into Phase II CF Patient Lung Function Preservation Trial

Continued on page 38

TILLMAN continued from page 37

Celtaxsys announced today that it has received a development award for $5 million from **Cystic Fibrosis Foundation Therapeutics, Inc.** (CFFT). The development award will help support a Phase II clinical trial of the company's lead development candidate, a once-daily, oral anti-inflammatory drug CTX-4430 in adults with CF. CTX-4430 is a selective inhibitor of Leukotriene A4 Hydrolase that targets re-balancing of a patient's over-activated inflammatory response without increasing susceptibility to infection. CTX-4430 has been granted orphan status for CF in both the US and the EU. CTX-4430 is a once-daily oral drug that is well tolerated. Importantly, in only two weeks in CF patients, CTX-4430 demonstrated positive results on well-established CF biomarkers, including encouraging reductions in sputum neutrophils. In addition, CTX-4430 demonstrated a statistically significant reduction versus placebo in neutrophil elastase, which has been shown to predict the onset of bronchiectasis and which can degrade CFTR and other protective proteins.
http://tinyurl.com/q5o34s4

FYI

Pseudomonas aeruginosa in CF and non-CF homes is found predominantly in drains. M.E. Purdy-Gibson, M. France, T.C. Hundley, N. Eid, S.K. Remold Journal of Cystic Fibrosis. May 2015. Volume 14, Issue 3, Pages 341–346

For patients with CF, Pseudomonas aeruginosa infection is a major contributor to progressive lung disease. While colonizing strains are thought to be primarily environmental, which environments are important in lung colonization is unclear. The findings from this study implicate drains as important potential sources of P. aeruginosa infection. They suggest that maximizing P. aeruginosa control efforts for drains would reduce exposure with minimal extra burden to CF patients and families.
http://tinyurl.com/q87q8e5

Inflammation in cystic fibrosis lung disease: Pathogenesis and therapy. Cantin AM, Hartl D, Konstan MW, Chmiel JF. **J Cyst Fibros. 2015 Mar 23**

This review examines the role of inflammation in the pathogenesis of CF lung disease, summarizes the results of past clinical trials and explores promising new anti-inflammatory options. The inflammation of the CF lung is dominated by neutrophils that release oxidants and proteases, particularly elastase. Neutrophil elastase in the CF airway secretions precedes the appearance of bronchiectasis, and correlates with lung function deterioration and respiratory exacerbations. Anti-inflammatory therapies are therefore of particular interest for CF lung disease but must be carefully studied to avoid suppressing critical elements of the inflammatory response and thus worsening infection.
http://tinyurl.com/qdv3j7r

Reduced bacterial colony count of anaerobic bacteria is associated with a worsening in lung clearance index and inflammation in cystic fibrosis. O'Neill K, Bradley JM, Johnston E, McGrath S, McIlreavey L, Rowan S, Reid A, Bradbury I, Einarsson G, Elborn JS, Tunney MM. PLoS One. 2015 May 20;10(5)

The aim of this study was to investigate the relationship between the colony count of aerobic and anaerobic bacteria, lung clearance index (LCI), spirometry and C-Reactive Protein (CRP) in patients with CF. The results indicate that lower abundance of aerobic and anaerobic bacteria may reflect microbiota disruption and disease progression in the CF lung.
http://tinyurl.com/qfjfw3g

TREATMENTS

Antibiotic management of methicillin-resistant Staphylococcus aureus-associated acute pulmonary exacerbations in cystic fibrosis. Fusco NM, Toussaint KA, Prescott WA Jr. **Ann Pharmacother. 2015 Apr;49(4):458-68**

The authors aimed to review the treatment of methicillin-resistant Staphylococcus aureus (MRSA)-associated acute pulmonary exacerbations (APEs) in CF. Data extrapolated from other populations suggest that vancomycin and linezolid are appropriate first-line treatment options for the treatment of APE secondary to MRSA. Second-line options include doxycycline or minocycline and trimethoprim/sulfamethoxazole, each of which may be useful in patients coinfected with other respiratory pathogens, for which they may provide overlapping coverage. Although potentially useful, clindamycin is limited by high rates of resistance, telavancin is limited by its toxicity profile, and tigecycline is limited by a lack of demonstrated efficacy for infections that are similar to those seen in the CF population.
http://tinyurl.com/ndnvp92

A Phase III, open-label, randomized trial to evaluate the safety and efficacy of levofloxacin inhalation solution (APT-1026) versus tobramycin inhalation solution in stable cystic fibrosis patients. Stuart Elborn J, Geller DE, Conrad D, Aaron SD, Smyth AR, Fischer R, Kerem E, Bell SC, Loutit JS, Dudley MN, Morgan EE, VanDevanter DR, Flume PA. J Cyst Fibros. 2015 Jan 12

The authors compared the safety and efficacy of LIS to tobramycin inhalation solution (TIS) in persons ≥12 years old with CF and chronic P. aeruginosa infection. They concluded that LIS is a safe and effective therapy for the management of CF patients with chronic Pseudomonas.
http://tinyurl.com/p66ufcx

Eradication of respiratory tract MRSA at a large adult cystic fibrosis centre. Hall H, Gadhok R, Alshafi K, Bilton D, Simmonds NJ. Respir Med. 2015

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Mar;109(3):357-63

The prevalence of MRSA in patients with CF is increasing. There is no consensus as to the optimum treatment. This study suggests that combined antibiotic therapy, particularly Rifampicin/Fusidic acid, is a well-tolerated and effective means of eradicating MRSA in patients with cystic fibrosis.
http://tinyurl.com/lv4vcg3

Randomized trial of efficacy and safety of dornase alfa delivered by eRapid nebulizer in cystic fibrosis patients. Sawicki GS, Chou W, Raimundo K, Trzaskoma B, Konstan MW. J Cyst Fibros. 2015 Apr 25.

Dornase alfa administered via jet nebulizer is indicated as a chronic respiratory medication for CF patients. Efficacy and safety of dornase alfa via an electronic nebulizer with vibrating membrane technology have not been formally assessed in randomized clinical trials. Administration of dornase alfa via the eRapid nebulizer resulted in comparable efficacy and safety, shorter nebulization times and higher patient preference.
http://tinyurl.com/ovgknln ▲

Laura Tillman is 67 and has CF. She is a former Director and President of USACFA. She and her husband, Lew, live in Northville, MI.

REMINDERS

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

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

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

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

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

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