CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
WINTER 2016

Letter From The President

By Jeanie Hanley, MD

In this Winter issue of *CF Roundtable,* the Focus topic is Dealing With Gastrointestinal Issues. We are fortunate to have Amy Braid discuss her experience and several of our columnists join in. If you haven't yet read Devin Wakefield's article on his GI challenges at work in the Autumn 2015 issue, you should. You will be entertained! Laura Tillman's "Information From The Internet" also highlights several important GI studies.

Regardless of the state of our lung disease, GI predicaments add a significant impact and can reduce our quality of life substantially. The gut functions to absorb and digest nutrients and expel waste. The GI tract covers a large area starting at the mouth through the throat and esophagus, to the stomach, small intestine (duodenum, jejunum and ileum) and large intestine (colon), ending with the rectum and anus. The pancreas, gall bladder and liver are within the abdomen and release enzymes and bile into the small intestine. As many of us know too well, there are a myriad of illnesses caused by CF on the gut from thrush, reflux, pancreatic insufficiency, abnormal bacteria in the gut (from too many antibiotics), gastroparesis, DIOS, diabetes etc.

> "Through this novel treatment of noninvasive manipulation of different areas of the body including the abdomen, they were able to help my abdomen work more efficiently."

Many of us have had colonoscopies (lower endoscopies) that evaluate the state of our lower GI (colon, rectum and anus) or upper endoscopies that check out our mouth to stomach and duodenal opening. The small intestine, a site of much CF pathology and at increased risk of cancer in CF, is not assessed with these screening exams. Only special tests will reveal if there's trouble. See Amy's article that describes her journey navigating another GI disease and the tests that helped, including many that looked specifically at the small intestine.

As for GI research articles, in "Information From The Internet" a new enzyme formulation for those on enteral feedings is introduced. Another study on glucose intolerance and how the severity affects our pulmonary function and clinical sta-

Continued on page 11

INSIDE THIS ISSUE

CF ROUNDTABLE
FOUNDED 1990
Vol. XXVI, No. 1

CF Roundtable (ISSN 1057-4220) is published quarterly by the United States Adult Cystic Fibrosis Association, Inc. (USACFA), a totally independent, 501(c)(3) tax exempt, nonprofit corporation whose Board of Directors all have CF. Articles in *CF Roundtable* may be reprinted only with advance written permission from USACFA. All submissions to *CF Roundtable* become the property of USACFA and should include the author's full name, address and phone number. Submissions are subject to editing as needed and we reserve the right to edit any comments that disparage another person either by name or situation. Requests for anonymity will be honored.

CF Roundtable now is free for everyone. Tax-deductible donations are gratefully accepted to help defray the costs of printing, production and mailing of the newsletter as well as the costs of website maintenance. Please subscribe online at: www.cfroundtable.com or by mailing in the subscription form at the back of this newsletter.

CF Roundtable does not give medical advice. Any medical opinions represented in these articles are those of the writer and do not represent the views of USACFA, any of our Sustaining Partners or any other group or individual. We strongly suggest you consult your doctor regarding any medical references and before altering your medical regimen in any way. USACFA does not endorse any products or procedures.

United States Adult Cystic Fibrosis Assn., Inc.
PO Box 1618
Gresham, OR 97030-0519
E-mail: cfroundtable@usacfa.org
www.cfroundtable.com

EDITOR'S NOTES

2016 is starting off with a bang for most of us. Some are having the warm weather that should have happened a few months ago. Others are fighting floods. Others are enduring bone-chilling cold. Whatever your weather, I hope you are able to stay comfortable and healthy.

We have both happy and sad news to report. The sad news is that columnist **Jennifer Hale** died in December. See her obituary on page 19. We will miss her positive outlook on life and her happy smile. The happy news is that we have given out more USACFA awards. At a first-ever special ceremony during the North American CF Conference in Phoenix, AZ, in October, **James Passamano** was presented the **USACFA Founders Award**. At the same ceremony, it was announced that **Isabel Stenzel Byrnes** was the recipient of the **Jacoby Angel Award**. You can read all about these and other presentations on page 22. Our congratulations to James and Isabel, well done.

Inside this issue **Beth Sufian** discusses questions about acquiring insurance and getting reasonable accommodation for disabilities in "Ask The Attorney." **Isabel Stenzel Byrnes**, in "Spirit Medicine," writes of the importance of love to our health and the important role our spouses play in our lives. **Lisa Cissell** reviews the book, *A Life In Men*, on page 20. "Parenting" finds **Colleen Veitengruber** telling of her journey to motherhood. The "Poetry Corner" has some of **Linda Stratton's** thoughts about hospitalization. As usual, **Laura Tillman** has done a fine job of compiling the latest "Information From The Internet." **Reid D'Amico** is the subject of "In The Spotlight." Also, he covers information about sinus clinical trials in "Searching For The Cure."

Be sure to read **Jeanie Hanley's** letter on the front page. It leads into our Focus topic, which is Dealing With Gastrointestinal Issues. **Amy Braid** writes of dealing with Lynch Syndrome as well as CF. **Andrea Eisenman** writes about her GI woes both pre- and post-transplant. In "Wellness" **Julie Desch** lists some ideas and tips for having a happy GI system. I continue the discussion in "Speeding Past 50" with some of my GI memories.

As we begin our 26th year of publishing *CF Roundtable*, I hope you will look at the upcoming Focus topics, which are listed on the opposite page. Perhaps one of them can give you an idea of something that you might like to write about. As always, you may write on any topic that relates to CF and is of interest to you. This is your publication and we want you to participate in it. We love photos of you, original CF-related cartoons and poetry, too.

I hope your 2016 is filled with good health, love and laughter.

Kathy

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - AbbVie, Boomer Esiason Foundation, Cystic Fibrosis Services/ Walgreens, Foundation Care, Gilead Sciences, Hill-Rom, Modern Health, and The Estate of Pamela Gordon Beaton; Pearl Sustaining Partners - Cystic Fibrosis Foundation; Diamond Sustaining Partners - Nancy Wech.

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Arch Biopartners Announce Successful Pre-Clinical Validation Studies For Pseudomonas Aeruginosa Treatment

Arch Biopartners Inc. recently announced that its drug candidate AB569, a novel potential drug agent that works as treatment for mucoid and non-mucoid Pseudomonas (P.) aeruginosa infections that are resistant to traditional antibiotics, has successfully concluded pre-clinical in vivo and in vitro validation trials. In all of the company's pre-clinical trials, the drug was found to be effective against P. aeruginosa. The company believes that these new pre-clinical findings provide scientific evidence to initiate a study in humans to assess the efficacy and safety of AB569 in patients with CF infected with P. aeruginosa.
http://tinyurl.com/pfhcupd

Five Genetic Regions Implicated in CF Severity

If a person has two faulty copies of the CFTR gene, he will have CF. But the severity of the disease will depend on environmental factors and many other genes. Locating these genes and altering their function could lead to new therapies. Now, a consortium of research institutions in the United States, Canada and France report that five regions of the human genome are home to the genetic variations that play major roles in CF disease severity. The International CF Modifier Consortium analyzed more than 8.5 million SNPs – or DNA sequence variations – over the span of 12 months to find the five genetic locations on chromosomes 3, 5, 6, 11, and X that were significantly associated with variation of lung disease. The consortium is now trying to link these genetic polymorphisms to gene expression – the amounts of proteins that genes create. This would allow them and other researchers to design more targeted therapies to lessen disease severity, decrease infections and hospitalizations and prolong life.
http://www.newswise.com/articles/view/640542/?sc=mwhn

Celtaxsys commences acebilustat Phase 2 trial in patients with CF in U.S.

Celtaxsys announced the commencement of the U.S. arm of an international Phase 2 clinical trial for its flagship compound, acebilustat, in patients with CF. The study will be con-

Continued on page 13

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**

Winter (Current) 2016: Dealing With Gastrointestinal Issues.

Spring (May) 2016: Managing Various Conditions (Under The CF Umbrella.) (Submissions due March 15, 2016.) Do you have conditions such as GERD, CF-related arthritis, blood pressure problems etc. that are not purely CF-related but you must manage under the umbrella of CF? Tell us about them and how you have dealt with them.

Summer (August) 2016: Living With Anticipatory Grief And Survivor's Guilt. (Submissions due June 15, 2016.) Do you live with anticipatory grief or survivor's guilt because of your CF or have you dealt with it in the past? How do/did you handle it? Please share any good suggestions you may have for managing these feelings.

Autumn (November) 2016: Advocates And Advocacy. (Submissions due September 15, 2016.)

ASK THE ATTORNEY

Answers To Readers' Questions

By Beth Sufian, JD

In the past three months readers have asked many questions about their rights in employment and education. This column will provide answers to some of the questions. The areas of education and employment are complex areas of the law. The following answers are a brief discussion of complex areas of law. Nothing in this article is meant to be legal advice about your specific situation but is meant to be only general information.

CF Roundtable *readers with questions about issues related to Social Security benefits, Medicare, Medicaid, health insurance coverage, legal rights in school and employment can contact the CF Legal Information Hotline at 1-800-622-0385 or CFLegal@sufianpassamano.com. The Hotline is sponsored by the CF Foundation and all calls are free of charge and confidential. The CF Legal Information Hotline can now schedule a specific time for you to speak with an attorney during the week or on the weekend.*

1. If I have CF, does an insurance company have to sell me a life insurance policy?

There are no federal laws that specifically require an insurance company to sell a person a life insurance policy regardless of health. Therefore, it is difficult if not impossible for a person with CF to purchase an individual life insurance policy.

Typically people with CF who have life insurance have enrolled as an employee benefit through their employer. Large employers often offer life insurance to employees, and the employee does not have to meet certain health criteria in order to be eligible for the policy. There are no laws that require an employer to provide life insurance coverage to employees.

Sometimes people with CF are able to purchase individual life insurance policies that will pay a benefit only if the person has died as a result of an accident.

2. I am in college and became sick right before final exams this semester. Do I have the right to take my final exams when I am feeling better? Or do I lose all the work I did in the past semester?

College students with CF often find themselves run-down and exhausted by the end of the school semester. Sometimes the student with CF gets sick and must either go into the hospital for treatment or receive treatment at home. It may become impossible to take the semester final exams. The Rehabilitation Act of 1973, Section 504, provides that entities that receive federal funds should not discriminate against a person who has a disability. Section 504 has been held to apply to students with disabilities who attend college and need reasonable accommodations in order to prevent discrimination. A student with CF who has a limitation in a major life activity, such as breathing, will most likely be protected from discrimination by the mandates of Section 504 of the Rehabilitation Act.

BETH SUFIAN

A student who meets the legal definition of a person with a disability and is enrolled at a college that receives federal funds can request a reasonable accommodation from the school. Almost all colleges and universities receive federal funds and so must follow the mandates of the Rehabilitation Act.

For example, if a student is sick at the end of the semester, the student may be able to obtain a reasonable accommodation of taking his final exams after the other students. The student would request the reasonable accommodation and then the school could give the student a grade of "Incomplete" in his courses. When the student recovers from his illness, he can take his final exams without penalty.

3. I work part-time and have asked my employer for a reasonable accommodation of being able to drink water while working as a cashier. The employer says he will not allow me to drink water.

Title I of the Americans with Disabilities Act (ADA) protects employees who have a disability if the employer has 15 or more employees. If the employee has a significant limitation in a major life activity, such as breathing, then the employee may have protection from discrimination under the ADA. An employee can request a reasonable accommodation if the accommodation is needed to perform the essential functions of the job.

Failure to provide a reasonable

accommodation to an employee who is protected under the ADA can be reported to the Equal Employment Opportunities Commission (EEOC) within 300 days of the act of discrimination. A person does not need to hire an attorney in order to file a charge of discrimination with the EEOC.

The EEOC will investigate the charge of discrimination. Sometimes the EEOC will try to resolve the complaint. Other times the EEOC will issue a "Right to Sue" letter. The employee then has 90 days to file a lawsuit in federal court. It often is very difficult to find an attorney who will bring a case under Title I of the ADA. If the employer is willing to mediate the charge of discrimination with the EEOC, a positive resolution may result. ▲

Beth is 50 and has CF. She is an attorney who specializes in disability law and is a Director of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues.

POETRY CORNER

LINDA STRATTON

Familiar Faces

By Linda Stratton

Held captive once again in a medical maze
of doctors, procedures and lung-cleansing care.
Weeks of illness culminate in this necessity,
giving myself over to inquisitive minds and healing hands.
Under the circumstance of infirmity it's strange,
to realize I'm pleased to see them again.
The nursing staff aware of my quirky needs,
from the timing of treatments to an extra blanket for bed.
There's warmth and safety in their knowing care,
becoming home away from home as CF rages on.
What a blessing it is greeting familiar faces,
with welcoming eyes of compassion and kindness.
Even through the haze of the illness inside me,
I treasure this aspect of my life as it is.

Linda is 61and has CF. She lives with and cares for her 88-year-old father in Denver, Colorado. A lover of cats, she also lives with Missy, a 13-year-old, Denver-bred Nebelung (specialty breed.) A lower altitude would be better, she acknowledges, but the beautiful change of seasons in the "mile high" city and love of family keeps her content. She also feels fortunate to be close to the CF Care Center at National Jewish Health.

SPIRIT MEDICINE

CF Partners: The Spirit Of Higher Love

By Isabel Stenzel Byrnes

'Tis the season of Valentine's Day, so I'd like to write about the capacity for loving, and how cystic fibrosis (CF) invites many of us to witness a higher form of love. Many adults with CF, like me, are granted the gift of companionship of a partner or spouse. Not everyone has a partner or spouse at this time, but my single CF friends remain optimistic. Or at least I hope they do.

At a CFRI conference many years ago, a father of a CF child approached Andrew, my husband, and told him, "You are the only one who is here voluntarily." He was right. I've been blessed to be married for almost 18 years — a length I never imagined possible back when I was a rather sick 26-year-old bride. Being a partner or spouse of a CF adult can take tremendous patience, compassion, sacrifice and tolerance. My husband has stayed in a job for the insurance; he has listened to my endless complaining about pain and other physical symptoms; he has spoken up for me when I was too out of it to assert myself; he has soothed my chronic anxiety of what else might go wrong with my body. And, so, in this article, I'd like to explore what makes someone fall in love with someone who has CF.

I never really dated much, but I did know from my youth that having CF would be a great weeding tool to find Mr. Right. A person who could handle my treatments, mucus, hospitalizations and uncertain lifespan would need to be mature and special. I didn't want to waste my time with the others who couldn't handle the CF life. But I did struggle with being unworthy of romantic love, because of the baggage of CF. Why would anyone want to put up with this? There are also so many "fish in the sea," of healthy, attractive women that men could choose from. Why go for me?

I have met many partners and spouses of adults with CF. They are an exceptional breed. I just attended a wedding of a friend with CF. As we were dancing, the bride, who does not have CF, commented that her husband went back to the hotel room, to use the bathroom, instead of using the one in the banquet hall. She gave me "that look," as if we understood a secret code. Yep. I get it. And how it's all okay. Since this issue is about gastrointestinal issues, I'd like to especially acknowledge the sainthood of our partners and spouses who can tolerate CF bathroom habits. I feel so sorry for my husband, but he takes it all in stride, even finding humor in my toxicity. I am so blessed.

Last month, I also went out to dinner with a newly transplanted friend, together with her husband. When the food arrived at the table, her husband immediately served us both from a communal dish, to reduce the risk of cross infection, with an unspoken but firm protectiveness towards his wife. I was touched by his instinct to keep her from harm.

ISABEL STENZEL BYRNES

I have many more stories, but in general, my impression is that CF partners and spouses collectively seem more patient, compassionate and self-sacrificing than the mainstream. Now, as I've gotten older, of course I acknowledge there are many reasons why people with CF would be loved. We have CF, but we're human! As crummy as CF can be, I also realize how blessed we are to have an illness where our personhood — who we really are — can still shine above and beyond the symptoms of this disease. And so we *can* be attractive — romantically, physically and emotionally — to others. And in the United States, people with CF are still very much in the pool of romantic candidates. In some cultures in Asia or the Middle East, we'd be off the table completely.

Maybe loving someone isn't a choice. Anthropologist Dr. Helen Fisher of Rutgers University, in her book, "Why We Love," explains the "indomitable, unquestionable and primordial human drive to love." She studies three types of romantic love: lust, attraction and attachment. Love is deeply embedded in the brain. She has found that love is a biological drive, more powerful than the sex drive. Lust, or the sex drive, makes us want to mate with anyone. Love is not the desire to mate, but to find one person to share life with — to form a "pair bond." In her research, she found that people in

love activate the ventral tegmental area (VTA) of the brain, which creates dopamine, a chemical connected to the reward system. This chemical spurs wanting, obsessiveness, risk-taking, motivation, focus, craving, separation anxiety, emotional dependence. Fisher shares that romantic love happens when one person takes on "special meaning." Attraction means there is "idealization and crystallization," where you forgive the faults and focus on what is adored. In couples who have been married for ten to twenty years and still report feeling "in love," the VTA in the brain still fires with deep attachment. Attachment happens when the partner and spouse share deep feelings of union with a long-term partner, in which they provide a sense of security and safety like no other. So, the point here is that we are wired to love. No matter what. And our partners and spouses happened to find us.

Besides mere biology, we've heard many phrases about the mysteries of love: "it was meant to be," "it was love at first sight" or "it's a match made in Heaven." Some people believe you fall in love with people you've loved from previous lives. Some say "God is love" and so God is made manifest when love is felt so intensely. Sometimes the chemistry of strangers just fits. It's baffling why love happens.

Back to why partners would be attracted to someone with CF, a friend once told me there is something profoundly romantic in loving someone who may be dying. All the best novels and dramas contain this theme; love on the edge is the most passionate love of all. But the ultimate price of loving is losing. My heart breaks every time I attend a funeral of a CF peer and see the devastation that the partner endures. But all the partners I've met have known of this possibility, yet still took the risk to love. I still remember one spouse who opened his eulogy to his wife with, "Seventeen years. That was more than I ever dreamed of."

In general, most partners would share that they don't regret loving someone with CF. Despite all the sacrifices, I realize now that my CF has given Andrew experiences that have enriched his character. My illness has opened his eyes to what truly matters. My limited lifespan has encouraged Andrew to live the best life he can possibly live, too. And my CF has offered Andrew opportunities to be part of vibrant communities and special events that he would have never been exposed to.

Of course I've benefited from learning and growing from Andrew's personality, hobby/professional communities and aspirations. Love is a two-way street, but I still think I gain more. There's no doubt about it; my husband's presence has literally kept me alive. First, Andrew spent ten years helping me with chest physical therapy. But there is a spiritual influence as well. There have been times when the CF struggle was so intense, I could have given up and released my fighting spirit. The question, "Who do you live for?" is so important for all of us to keep up our drive to be compliant and hopeful. And sadly, I've known a number of friends who have died after a breakup or divorce. What could be more meaningful to a person than to be in a relationship with someone where your mere presence is life-prolonging and life-affirming? As C. S. Lewis wrote, "Love is not affectionate feeling but a steady wish for the loved person's ultimate good as far as it can be obtained."

Partners and spouses play another important role in lifting our spirits. I've lately witnessed a spouse tell a CF friend, his wife, that she needs to get a job. Later I asked her how she felt about his directive, and she shared that she was glad she had someone to push her, to nudge her, to encourage her. Our closest partners can be blatantly honest, sometimes even judgmental or critical, but in healthy relationships this can be all done with good intentions — for us to live the best lives we are capable of. It sure feels good to be so open with someone, and know that they still love you.

I don't mean to be idealistic here, because relationships are hard work. Conflict is common, egos clash, blurred boundaries mean we say and do things that hurt the other person. Relationships are exhausting, when energy is limited anyway. CF takes so much time, how are we supposed to devote more time to our relationships? Relationships are a graceful dance between self and other, me versus us, togetherness and privacy, giving and receiving. They are dynamic and need constant attention. Some relationships have their time to begin and end. For some, sadly, CF *is* too much. The expectations of life are not met when CF is in the picture. This is the heavy cost of being in a relationship. The alternative is closing the heart, living in solitude or loneliness or putting oneself back on the market and opening to new love.

Most importantly, our partners bear witness to our lives with CF. They walk alongside us, seeing what this roller coaster ride is really like. My favorite Chinese proverb is, "Joy shared is joy doubled, and sorrow shared is sorrow halved." Ain't that the truth? Just the fact that we can share this journey with someone who chooses this wild ride is mind-blowing. So, my hat is off to all the partners and spouses out there. Thank you for your courage, strength and love. If more of this type of love were cultivated, our world would be a better place. ▲

Isabel is 44 and has CF. She lives in Redwood City, CA. She works as a bereavement counselor. You may contact her at: Isabear27@hotmail.com

SPEEDING PAST 50

GI Memories

By Kathy Russell

Some of my earliest memories have to do with my GI (gastrointestinal) system. When I was a little girl, it was common to have potty chairs for children. I had one. I remember sitting on it for what seemed like hours at a time. It was as if I always felt that I had "to go." I would sit there and read, or at least look at, books. I spent a lot of time rocking back and forth. My mother tried various foods and medicaments that were to help alleviate constipation; although, to be perfectly honest, I wasn't really constipated. I had very large stools; sometimes several a day. None of the methods that mother tried seemed to be very effective.

I always had a "pot belly." Even when I was a skinny little thing with spindly arms and legs, my belly was sticking out as if I were an old beer drinker. At five or seven, I didn't mind having a big belly. By the time I was a teenager it really bothered me. There were times that I looked as if I were eight months pregnant when, really, I was underweight. Now that I'm not underweight I still have the belly.

I was unaware of some of my GI problems until I started having rectal bleeding. That captured my attention, quickly. I went to one of the docs with whom I worked. I knew that he was a great rectal surgeon. I trusted him and his assessment of my troubles. He said I had a severe rectal prolapse with bad hemorrhoids. I had surgery that repaired all the problems.

Of course, there is one problem that doesn't seem to change much. It is odor. Our stools and flatulence or, as most of us know it, *gas* are prone to be terrible odors. For that reason I don't usually use a bathroom away from home. Also, I am very careful about not eating foods that cause gas when I am going to be around others.

I took enzymes for quite a while and found that I needed to take increasingly larger doses. The docs kept telling me that it was okay to take larger and larger doses. This didn't make sense to me. I was concerned about residual damage from taking such large doses of these meds. Years later, in studies it was discovered that large doses of enzymes could cause fibrosing colonopathy. I'm glad that I balked at those large doses.

> *"Because of my low fat choices, I am able to function quite well without taking enzymes."*

KATHY RUSSELL

I always have eaten a low fat diet. This is my choice. Fat has never felt good in my mouth. Some fats cause a fiery pain in my esophagus. If I get even a small amount of one of those fats into my throat, it will feel as if I had poured lighter fluid down my throat and had lit it on fire. It is incredibly nasty pain. Because of my low fat choices, I am able to function quite well without taking enzymes.

My life has been a series of taking enzymes for a few years, not taking them for a few more, then taking them again and, finally, not taking them for about the past 20 years. I feel that I am able to tell if I need to think of taking them again. So far, so good.

Along with taking or not taking enzymes, I have had bouts of DIOS (distal intestinal obstructive syndrome). I have been hospitalized a few times because of DIOS, although usually I am able to get it to move without medical intervention. My DIOS seems to be caused by a very lazy gut. It is as if my gut just decides to take a vacation and not work for a while. It loves to "clamp down" and not do anything. When one doc did a colonoscopy on me, he said that he never had seen anyone's colon clamp down the way mine did. It was impossible to pass the scope until he had given me more than twice the normal dose of med to relax my muscles. He said that he never wanted to go through that again.

I agree with him. I paid for that high dosage by having a terrible headache and not being able to do even the simplest arithmetic problems for several days afterward. I also had mild

amnesia. It scared me enough that I have made sure that I didn't have a repeat incident.

I went through a time of many years where I would have intermittent bad gut pain. It might stop me cold as I was going about my daily activities or it would wake me from a sound sleep. The pain would be so severe that I found relief only by getting onto my elbows and knees with a hot water bottle or heating pad tucked up against my abdomen. It might take an hour or more for the pain to ease.

No one was able to identify the cause of those pains, although sometimes they thought it might be my diverticulosis acting up. I had CT scans and MRIs without any diagnosis being made. There were several conflicting ideas of the cause, but no one really knew.

About 14 years ago, I was experiencing such a sharp pain in my low abdomen that I couldn't even zip my trousers. I couldn't eat and felt miserable. I went to the emergency department and they thought that my bladder was enlarged. That is how it looked on X-rays. Then they did a CT scan and discovered that I had a large abscess on my colon. After two weeks in the hospital on IVs and no food, I still had a stool blockage.

I had a hemicolectomy. The evening before surgery, the abscess ruptured. Fortunately for me, it ruptured into the bowel rather than the abdomen. I had been on antibiotics for two weeks and sulfa for a few days prior to surgery so I had no problems with post-op infections. Getting rid of that half of my colon was a good thing for me. I haven't missed it and I don't miss the pain it used to cause. (And just for the record, yes, I now have a semi-colon!)

I went along okay for a few years. Then I got another blockage. This time it was my own fault. I did something stupid and I paid for it. I know that I have to be careful about not eating too much dry food without taking in plenty of liquids with it. Also, I know that I should be cautious about eating nuts. Well, I wasn't. I had some really tasty, dry roasted peanuts. The salt on them kept enticing me to have "just a few more." Of course, I had too many more. I got a lump of peanuts stuck at the end of my small intestine.

Another trip to the emergency department ensued. I was admitted and the doc and I tried to get the blockage to move. After several days with no success, it was back to surgery. As it turned out, not only was there a blockage, but my small intestine was twisted. The doc moved the blockage manually and untwisted my intestine. He said that everything in my abdomen was attached to everything else by adhesions. He spent a long time getting rid of all the adhesions. Such fun. Not.

Since that surgery, I have been taking polyethylene glycol (Miralax equivalent) every day. I do 17 grams of the powder dissolved in diluted Newman's Own Limeade. (I have to dilute it because it is too sweet for me.) The lime covers the taste and feel of the powder and it is not bad at all. As long as I take it, faithfully, I am fine. It is so nice to have something work as it does for other people.

I have had GERD (gastroesophageal reflux disease) for as long as I can remember. When I would lie down, acid would roll right back up my throat. That was both unpleasant and painful. Nothing seemed to help it for very long. Then a doc had me try Nexium. Wahoo! Relief! I have been taking esomeprazole magnesium, which is the generic of Nexium, for several years and I no longer suffer with GERD.

Now, thanks to polyethylene glycol and esomeprazole magnesium, I am feeling quite well. I still am very careful about what I eat. I avoid most fats. I have found that sunflower oil and good olive oil are okay for most of my cooking needs. I can use butter, occasionally, for added flavor. I am careful to not use too much butter, though. I eat fish and chicken more than other types of meat. I eat red meats only rarely. I eat lots of fruits and vegetables. I like them as natural as possible. No gravies or sauces for me. I also eat a lot of grains. I believe that these practices help me stay healthier. Since I am nearly 72 and I am still surviving quite well with CF, maybe my diet works okay.

If you suffer with GI problems, I hope that you are able to find the proper mix of diet, meds and treatments to take care of your troubles and to allow you to live comfortably.

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

In Memory

Lisa Marie Wilder, 49
Piggott, AR
Died on October 25, 2015

Jennifer Hale, 43
St. Petersburg, FL
Died on December 5, 2015

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

WELLNESS

The Care And Feeding Of The CF Digestive Tract

By Julie Desch, MD

The title above is sort of tongue in cheek. After 55 years of stomachaches and digestive misadventures, it is not like I'm the expert. I know only (sort of) what NOT to do, after 55 years of experimentation. Additionally, just like almost any other "fact" you hear about CF in its varied renditions, results are variable. What worked disastrously for me might be your secret to a long and happy life. What keeps me pooping happily might stop you up for weeks. Read with caution.

A brief disclaimer: I do not have personal experience with one of the biggest gastrointestinal tract challenges, CFRD. The pancreas, as a very important organ in the GI tract, serves two functions. The exocrine function of providing digestive enzymes is one you may be intimately familiar with, as I am. The endocrine function is another animal with which I have had little experience (other than the time I almost killed myself with an overdose of insulin). Therefore, I am going to let others with much more experience than I have discuss CFRD.

The following is a list of experiments that I have attempted and suggest you DO NOT undertake:

1) Decide that enzymes are not for you (assuming you are pancreatic insufficient like me) without also diminishing your fat intake drastically. Of course, any CF caregiver will tell you NOT to diminish your fat intake at all, but just take your damn pills. As a kid, I refused the pills, yet continued eating Kentucky Fried Chicken and Long John Silver's fish and chips. Hence, the daily stomachaches. Thinking back, I would wager that at least a quarter of my formative years were spent on the toilet, doubled over in pain. Once I figured out that a healthy low fat diet cured my stomachaches without requiring enzymes, I was golden for almost 20 years. Now, as I believe I have finally lost the last acinus of pancreatic exocrine cells, I resort to enzymes. I am currently in love with butter.

2) Think, "Because you can eat pretty much anything and not gain weight," that what you put in your mouth doesn't have significant effects on your body and health in general. This was another folly of my youth. It most certainly does. Foods matter. Pouring crap into your system (refined carbs, sugars, soda, fats that are created in a lab) can turn your body into an inflamed mess. Eating "clean" is not just for CFTR-able people. We benefit as well. I'm now an avid reader about something termed "epigenetics." Basically, epigenetic factors control how genes are expressed. I always wondered why my genes, which should be buried in the ground right now if you read the older research, are alive and kicking while my siblings, who had—one would think—the same genetic defects, suffered much more than I have and eventually died from their disease. Epigenetic factors include *what you put into your body.* I'm sure other factors are involved as well, like modifier genes and environmental exposures. My point is that you have more control than you think when you realize that what you eat and drink could affect how your genes are expressed. Choose wisely.

> *"Pouring crap into your system (refined carbs, sugars, soda, fats that are created in a lab) can turn your body into an inflamed mess."*

JULIE DESCH, MD

3) Think that hemorrhoids will just "disappear" someday. After years and years of straining to cough, straining to poop, sitting on the toilet at the mercy of gravity for hours etc., these little critters are bound to appear. They must be destroyed. Make friends with your local general surgeon and her banding gun (or whatever it is called). Then, make friends with your ice pack.

Here are some of my favorite digestive tract tips:

1) Take probiotics. I favor

Culturelle. Think about the load of antibiotics you pour into your system on a daily basis. There is an ongoing assault to all bacteria, oral and inhaled, chronically and the intermittent IV drugs during exacerbations. They kill all bacteria, the good and the bad, and this is not a good thing when it comes to the colon, where beneficial bacteria are needed to help digest food and even regulate the immune system. If you don't want to take probiotic pills, you can eat yogurt, kefir, miso, tempeh, kombucha or kimchi. These all contain the same "good bacteria" that go missing after antibiotic use. As an aside, did you know that you have more bacteria on and in your person than you have cells in your body? Freaky.

2) Drink water: What does this have to do with the digestive tract? Well, let's think about the last time you were "plugged up," so to speak. Did you take some Miralax? The way Miralax and its big brother, GoLytely, work is by osmosis. The active ingredient, polyethylene glycol, essentially pulls water from the cells lining the digestive tract into the lumen, where the plugged up poop clings tenaciously to the gut lining. This water helps move things along. So water is key to a good poop. Adequate hydration is also a good plan for the lungs, by the way. Don't skimp on water.

3) Walk daily. Walking and water are two of the best catalysts to regular bowel movements, right up there with morning caffeine. Thinking back, the times when constipation becomes the biggest problem for me are almost always during hospital stays. What is missing the most in the hospital (especially when I really feel crappy)? Moving! I lie in the bed, day and night, moving only when I need to go to the bathroom, which is sadly, almost never. Usually, as soon as I feel well enough to walk again, my morning ritual resumes and life is good.

4) The liver, like the pancreas, is part of the digestive tract. It creates bile, which is important for the digestion of fats. If you are like me and many others with CF, you have had your gallbladder (the storage unit for bile) surgically removed because the bile we form, instead of the normal thin green fluid, is thick green sludge which has a tendency to form stones. In addition to gallbladder disease, this can cause the cells of the liver to be crabby as well. They have a hard life, so I recommend that you treat them with care. Long story short, be careful about alcohol intake. Alcohol is, after all, a poison that must be metabolized by the liver. If you overtax the liver with heavy alcohol use on top of the normal stress it deals with due to weird, thick bile that tends to back up, you are asking for liver damage. Alcohol is also dehydrating (see point 2 above).

These are just a few of my favorite do's and don'ts regarding the GI tract. I'm sure you can think of thousands of equally important ideas. GI tracts and how we love them are very personal. I wish you all luck with that very important relationship. ▲

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

HANLEY continued from page 1

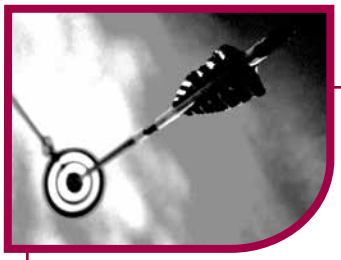
tus is considered. On our *CF Roundtable* blog from December 29, 2015, we listed an important study on the effects of alcohol on CFTR and the development of pancreatitis. A definite must-read. There are many more described on our blog. I hope that you subscribe to our blog so that you don't miss any of the latest research.

There are also clinical trials that involve the gut. I recently participated in a small study in Northern California undergoing osteopathic manipulative treatment. Please see Sara Modlin's summary article of this study in the Winter CFRI newsletter (www.cfri.org). Through this novel treatment of noninvasive manipulation of different areas of the body including the abdomen, they were able to help my abdomen work more efficiently, with less bloating and other benefits. I never would have known about this type of treatment without participating in this clinical trial. This is my shout out to encourage all those with CF to participate in clinical trials in your area whether GI, respiratory or other. It is quite a learning experience.

I'm reminded of a joke of how important the GI viscera are. Here it goes: different organs (brain, heart etc.) of the body were discussing who the boss of the body is. They all had valid reasons for being in charge – the brain, the blood etc. When the anus said, "I am the boss!" all the organs laughed at it. It was so mad to be laughed at that it clamped down shut. After three days, all the other areas of the body were pretty miserable and conceded that indeed the a$$@%!! is in charge.

I hope you enjoy this issue of *CF Roundtable* and are having a healthy new year. ▲

Jeanie Hanley is 53 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.



FOCUS TOPIC

DEALING WITH GASTROINTESTINAL ISSUES

Lynch Syndrome And Cystic Fibrosis: My Life With Both Genetic Diseases

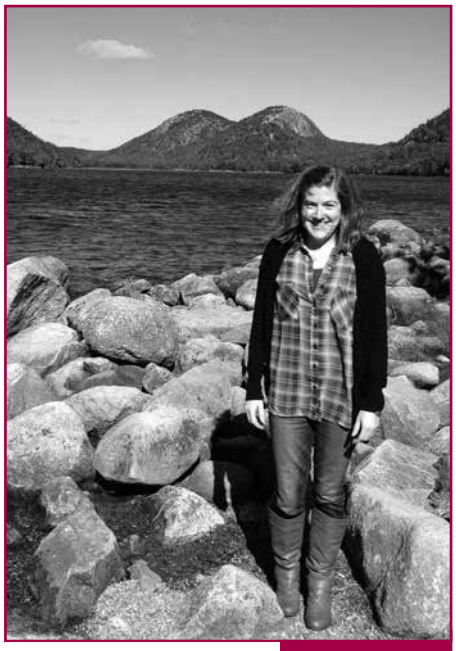
By Amy Braid

What is Lynch Syndrome? Lynch is much easier to say than hereditary nonpolyposis colorectal cancer, or HNPCC for short. The gist of it is a predisposition to certain types of cancers. For my mutation (hMLH1) they include the colon and rectum, stomach, small intestine, liver, gallbladder, bile ducts, upper urinary tract, brain, skin, female reproductive organs and more recent studies have shown that there is an increased risk of pancreatic cancer. In order for a family to be considered to have Lynch, there have to be two successive generations of either colon cancer, or colon polyps and colon cancer. For my family it is every generation with both cancer and polyps, and multiples in that generation. My father was diagnosed at the youngest age in my family, 26. The family history is striking – my father, his mother, three of her brothers, her parents, one of my father's cousins, five of my father's sisters and his brother (not all are colon cancer, some are reproductive organs and other GI organs, too). And this is only what we know of. As for the current generation that my brother and I are a part of, no one has been diagnosed, though some of my cousins have been tested or they just get colonoscopies every few years. My brother was tested and found to not be a carrier. Good news for his children!

My diagnosis:

At 26 years old my father was diagnosed with colon cancer. My mother had just given birth to me and they were only a few years into their marriage. My father was not given a good chance to survive. My mother feared she would be raising me alone. Miraculously my father survived, and he is still here today. Because of this scare, my younger brother and I were aware of our chances of developing colon cancer at a young age. We knew the family history was very strong. I remember being young and hearing "genetic cancer." I had no idea what that meant except that it ran in my family.

The day my CF doctor and I decided I needed to be tested for the gene was a day I will never forget. I had been having some issues with my bowels and was worried because I was past the age my father was when he had had cancer. I mentioned it to my then-CF



AMY BRAID

doctor. He replied that coughing could cause hemorrhoids and not to be worried about small amounts of blood in the stool. Then I told him my family history. He was on the phone with the genetic department at the University of Pennsylvania immediately. He wanted me seen as soon as possible. A few weeks later, in late June of 2009, I sat patiently in the waiting room of the genetic counsellor's office. I was not sure what to expect, though I was feeling pretty confident that I would come back positive for Lynch. She and I went over the detailed family history I had brought with me. We joked around and I told her I was positive I had the gene and that my brother was clear. We said our goodbyes and I expected to hear from her in a few weeks. About two weeks later she called me with the news I had been expecting: hMLH1 mutation for Lynch Syndrome. As much as I had been expecting the diagnosis, I was devastated. It is one thing to think you may have the gene, it is another to be told you definitely do.

Within a few weeks I had my first of many colonoscopies and just in the nick of time. Some of the polyps they removed were a little too developed for anyone's liking. They were removed with no issues and I avoided following in my family's footsteps.

My cancer scare:

Shortly after I was evaluated for bilateral lung transplant in 2011, I was told I needed to get an MRI of my abdomen done. There was a cyst on my pancreas that was larger than the doctors liked and they wanted it to be monitored. So in I went for my first of what would be many MRIs. Within a year it was thought that the cyst had more than doubled in size. I say thought because the doctors were unsure of the original size. During one of my yearly colonoscopies they tried to get a biopsy of the cyst but were unable to, so instead settled for an endoscopic ultrasound. This again showed that the cyst was larger than they would like and

had grown. The cyst was still under the 3cm "rule of thumb," so the plan was to monitor it. However, in late 2013 during my yearly colonoscopy, the doctor was finally able to get a biopsy of the cyst. The results were alarming. The levels they use to determine if it is cancerous were elevated to a pre-cancerous state. I was referred to a pancreatic surgeon to discuss my options. My personal opinion was to get the thing out of me no matter what.

I met with the surgeon and his team in early 2014. We discussed surgery and all that it entailed. It would be more invasive than I had thought, requiring the removal of my gall bladder, parts of the bile ducts and the head of my pancreas. However, I was still considering moving forward with the surgery. In the meantime, the surgeon contacted the transplant team and my CF doctor to discuss the results of the MRIs and the biopsy. The collaborative consensus was I needed to have the cyst removed to determine if it was pre-cancerous or only a cyst related to cystic fibrosis. So in July 2014 I was wheeled into surgery and had the Whipple procedure done. I passed with flying colors and the cyst was ruled a CF cyst.

My Lynch and CF dynamic:

Lynch affects my cystic fibrosis in many ways. I need to have yearly testing done to be sure I am not growing any cancerous cells in my body. Currently I get colonoscopies, a thyroid ultrasound, an upper endoscopy, a skin checkup and an MRI of my pancreas even after having surgery. Having CF complicates my colonoscopies and upper endoscopies. A prep for someone without CF takes only the day before the procedure. If the procedure is on Thursday they stop eating Tuesday night, clean out Wednesday and are nice and clean for Thursday. Not for me. I need to stop eating Saturday night and eat clear liquids/foods on Sunday. Then Monday, Tuesday and Wednesday I have to clean out. And even with this I am not always "perfect" but good enough to get good results. I have devised my own prep and have given it to many of my CF friends who need to have colonoscopies done.

Lynch has also played a significant role in my evaluation for bilateral lung transplant. The team at my transplant center was not sure what to do with me. They had many meetings and in the end decided I would be a good candidate, under the strict terms that once I am transplanted, those yearly tests become bi-yearly tests. Since there is a much higher risk for cancer post-transplant because of the immuno-suppressants, my risk will be even greater because of my genetic mutation. As it stands now with my yearly colonoscopies, there are always a few adenomatous polyps removed (these polyps when left to fester can turn into cancer). In 2011 I had a full hysterectomy done because of Lynch. Many years of reproductive issues coupled with my desire to not pass my genes on to children led me to have all my reproductive organs removed. It was also easier to take the organs out than to have to worry about them post-transplant. Furthermore, the Whipple procedure was done because the transplant team did not want to take the added risk that I had precancerous cysts on my pancreas. They removed me from the list until everything could be figured out. And the Whipple was the only way to be absolutely certain one way or the other what the cyst was. ▲

Amy is 34 and has CF. She lives outside of Boston with her husband and two step-children. You may contact her at amysilcox80@gmail.com.

TILLMAN continued from page 3

ducted at approximately 60 sites in the United States and European Union, with the EU sites anticipated to begin enrollment early 2016. This landmark clinical trial testing once daily oral acebilustat treatment over 48 weeks could be the first to establish proof-of-concept for an anti-inflammatory treatment specifically designed to prevent long-term loss of lung function in CF patients. The study will test once daily oral doses of 50 mg and 100 mg acebilustat against placebo. Acebilustat is a first-in-class neutrophil modulator that controls a key inflammatory signal over-expressed in CF. Acebilustat has been granted orphan drug status for the treatment of CF in the U.S. and the EU. Results from initial clinical studies in CF patients demonstrated acebilustat's ability to moderate the over-activated inflammatory response in CF, decreasing neutrophils in the lung by 65% and decreasing damaging neutrophil elastase in just two weeks of treatment. Acebilustat did this without jeopardizing the patient's immune response to infection. Importantly, acebilustat treatment is applicable to all CF patients irrespective of their gene mutation.

http://tinyurl.com/zodh76g

Phase 1b Results for N91115 CF Drug Presented at the North American CF Conference

Nivalis Therapeutics, Inc., reports positive results from a Phase 1b clinical study evaluating its CF drug candidate N91115, a novel stabilizer of the CF transmembrane conductance regulator (CFTR) protein. The clinical trial demonstrated favorable safety, tolerability and pharmacokinetics in adult CF patients with two copies of the F508del-

Continued on page 21

GI Woes—Pre- And Post-Transplant

By Andrea Eisenman

It is said that you are what you eat. Not a pretty picture because it used to be that I could, and would, eat everything without any adverse reactions from my stomach. I took enzymes, of course, but it was like my stomach was an iron drum, able to withstand any assault I threw at it.

Now, that is not to say I didn't have to "go" many times a day. Which brought its own form of humiliation due to the stench and time it took me in the bathroom. It was a running joke to those of my close friends who knew I could "destroy" a bathroom and therefore were warned to go before me or forever hold their, you know, whatever they had to do in there. But I never had gut-wrenching pains or cramps after I was about 10 or 11 years old. I think those pains may have been due to me not taking my enzymes, at that time Viokase, when I was younger. I was not a compliant lass.

That all changed post-transplant. Or, it started slowly. Things I could eat without mishap became off limits. Random things such as pomegranates became instant diarrhea. (I know, TMI.) And then on doctors' orders, I was not allowed to eat sushi and raw shellfish, which I loved and had eaten all my life. I started to react to bacteria on the fish more than before. And the "outcome" was not good. So, no more raw fish or even cooked shellfish such as clams and mussels. Too risky. If there was just one that was bad, I was very sick. I learned after this happened once or twice.

Another change came after the removal of my gall bladder, five years after transplant. It seems I had developed a mass in my gall bladder and because of risk factors and my having diabetes, it had to come out. Fatty food like ribs or certain cuts of meat entailed taking many more enzymes. But I was not going to give up everything. I still wanted to think that I could eat almost anything.

Just recently, I started to develop unbearable acid reflux. Because I take immuno-suppressants for my donated lungs, I need to also take antacid medications. While I have breakfast, I take my morning vitamins and with them a proton-pump inhibitor – Pantoprazole. Then two hours later, I take prednisone plus the immune-suppressant, cyclosporine. Recently, my cyclosporine was readjusted and I had to take a bit more to have the correct level in my system. I really didn't think the reflux had to do with that, but I could not get rid of the reflux.

ANDREA EISENMAN

> "So, while trying to solve my acid reflux, another problem emerged."

The reflux was plaguing me all day long—when I woke, when I went to sleep at night. It felt like fire and burned non-stop just below my throat. I tried to curb acidic foods, but that didn't help. Then I started taking Pantoprazole twice day. That didn't work. Then Pepcid was added at night when I took my second dose of cyclosporine. Again, it didn't work.

It was confounding and very uncomfortable. I was working with my doctor and the nurse coordinators at my transplant clinic. They grew concerned because it is not just the pain, as you may know, but the risk of rejection of my transplanted lungs that they feared. As the acid snakes its way up the esophagus, it can go into one's lungs and this can cause rejection. I was sent for some testing. I went through a gastric emptying test, which was normal, and a swallow test that showed acid reflux. The reflux was not severe enough for me to be a candidate for a Nissen Fundoplication procedure, for which I was thankful. But the acid was still a concern. Finally, it started to subside. And I am not exactly sure why.

Maybe because I was on so much antacid medication, I had no stomach acid whatsoever. This was probably the case because my digestion became so

bad all of a sudden. What I used to eat with one or two Creon 24s was no longer being processed very well. After seeing a CF doctor for something else, I talked to her about how my digestion had changed for the worse. It was recommended that I start to use more Creon 24s and, for my weight, it was recommended I double the dose. It has helped tremendously. While constipation has never been my problem except after taking pain medication post-surgery, I was worried about getting a blockage. The doctor assured me this was doubtful with the recommended dosage of Creons.

So, while trying to solve my acid reflux, another problem emerged. I never really thought about this but our stomach acid is there for a good reason, to help us digest our food. It made me realize how delicate a balance people with CF and transplant have to live by, or at least I do.

For now, my reflux has receded. I am starting to back off of taking Pantoprazole twice a day and using Pepcid at night. So far, it is working. To aid my digestion, I also added in some probiotics that are taken three times a day. All in an effort to get my GI flora back into good balance, to support a healthier me.

Now I am gentler and kinder to my gut. I watch more closely what I eat to avoid really fatty stuff or things that may cause my stomach to feel unsettled while trying not to stuff my face and eat just before bedtime. So far though, I have not learned what makes me bloat up like an iron drum. And I may never. But for now, my GI tract and digestion are much improved. ▲

Andrea is 51 and has CF. She is a Director of USACFA and is Webmaster and Executive Editor of CF Roundtable. *Her contact information is on page 2.*

Mailbox

Please accept this donation in loving memory of my son, Thaddeus Novak, who died on November 1, 2014, at age 34, following a bilateral lung transplant. I have been an avid reader of *CF Roundtable* for many years. Reading so many accounts straight from the trenches was sometimes painful and scary but often helpful and encouraging. Thanks for what you do.

Janet King
Tenafly, NJ

Keep up the good work!

James Yankaskas
Chapel Hill, NC

I live alone with CF. I'm declining now – so it's a struggle.

Jerome F. Lape, Jr.
Mishawaka, IN

Thanks for a terrific publication!

Tina Dimick
Ansonia, CT

I appreciated today's blog post. Jimmy was my uncle and CF role model in the '80s/'90s. Orkambi (etc.) didn't make it in time for him, but I'm reaping the benefits and think that would make him happy.

Janice Buck
Arlington, VA

I love reading your publication. Thank you!

Sara Kominsky
Blacksburg, VA

Enclosed is a donation in memory of Amy Young. She lost her battle with CF on January 3, 2015. Amy was in the hospital when my daughter, Ginnie, had her first hospital stay with CF at age 13. Amy was a true inspiration for all of us. We learned during our stay that she was a "Christmas" baby. Holiday babies often are forgotten in the celebrations. Ginnie and Amy kept in touch, and I always sent her a birthday card and gift. I always reminded her how special she was, because she shared a birthday with Jesus.

Amy turned 50 in 2014 and Ginnie turned 38 this year. Amy sent me my first subscription for *CF Roundtable*. It has been a great source of hope, information and healing.

Please continue the good works and know that all of you are in my prayers. Merry Christmas and Happy New Year!

Nina Ferrell
Elizabethtown, IL

Thank you for all of the great information!

Linda Buyaskas
Redwood City, CA

Many thanks for the years of *CF Roundtable* we have received. While our daughter with CF was a child and a teen, we read it for motivation and inspiration. Now that she is 25, it means even more to us.

Wishing you all every blessing in 2016.

Thomas & Maureen Marlow
Staten Island, NY

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

Michelle Allen
Portland, OR
64 on September 14, 2015

Jerome F. Lape, Jr.
Mishawaka, IN
69 on October 11, 2015

David Versteeg
Via e-mail from The Netherlands
51 on December 12, 2015

Lois Weisenbacher
Riverhead, NY
69 on December 14, 2015

NEW BEGINNINGS

Married

Carl Robinson and Kara McCann
On November 22, 2015
On Asilomar Beach, Pacific Grove, CA

THROUGH THE LOOKING GLASS

PHOTO BY STEPHEN BOYER

Pill

You wanna come inside my mouth?
You dirty mind,
my tongue is dirtier. these pills
are bitter. Gelatin capsules really
aren't that scary...
How's the weather out there?
How are your kids? Your life?
Your dog? What color are
my teeth?

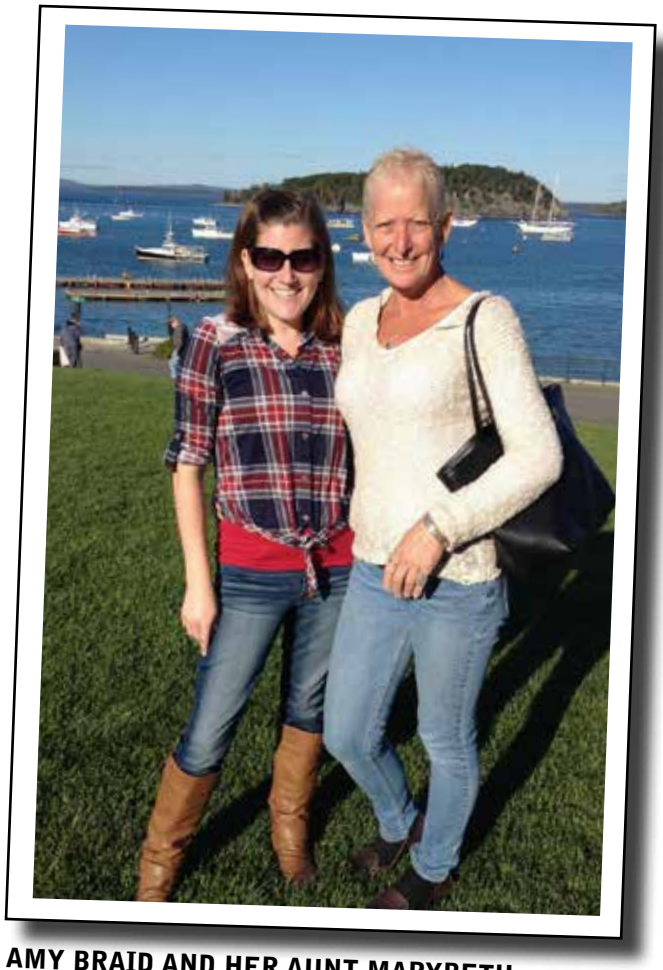
What is the
color of humility?
man, I'm bored. let's Keep
this train moving. move on
to a more interesting denial.
If I close my teeth and
grin, do these pills disappear?

Then quit pretending.

-Todd G., 2002

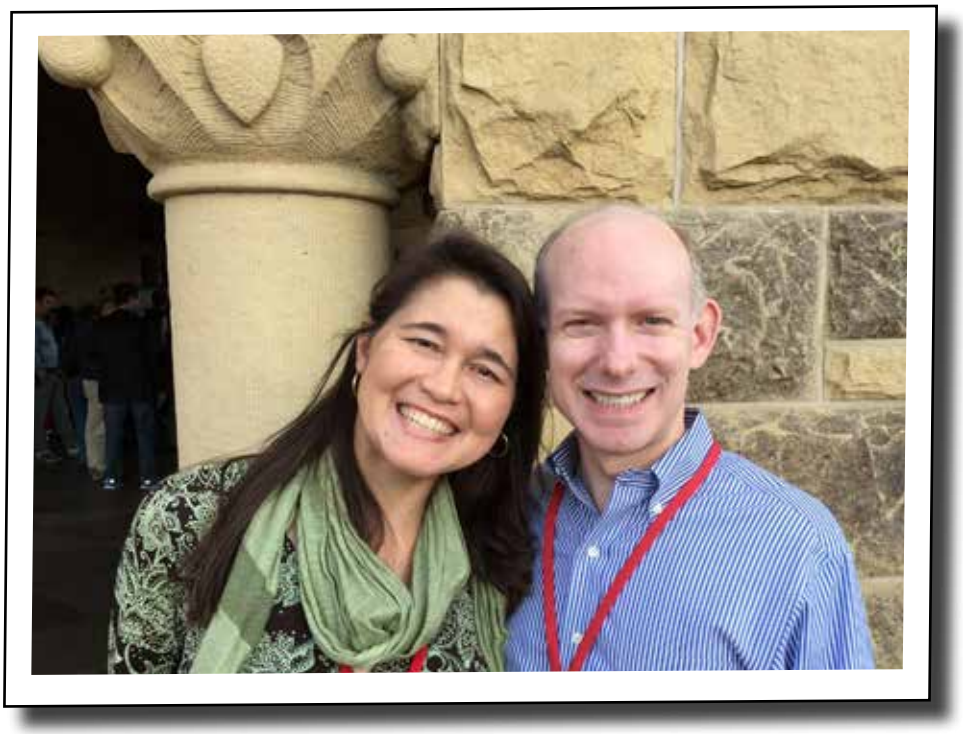
"Through the Looking Glass: Images of Adults with Cystic Fibrosis" and "Caregiver Stories" are projects of Breathing Room, a non-profit organization. Breathing Room hosts these and other projects to facilitate open and candid communication in the CF community, supports the development of a community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

FROM OUR FAMILY PHOTO ALBUM...



AMY BRAID AND HER AUNT MARYBETH GRAMIAK.

VEITENGRUBER FAMILY CHRISTMAS PHOTO – COLLEEN, TIM AND ANNA (15 MONTHS).



ISA STENZEL BYRNES AND ANDREW BYRNES.

DAD (JIM GROSSHANS) AND LINDA STRATTON ON THEIR WEEKLY LUNCHEON DATE, OCTOBER 2015.

ANDREA EISENMAN CHILLS OUT IN PUERTO RICO IN DECEMBER 2015.

Columnist And Former Director Of USACFA – Jennifer Hale

January 29, 1972 - December 5, 2015

Jennifer Hale, who was a USACFA Director in 2012 and 2013, wrote what she knew. In her *CF Roundtable* column, she discussed what she went through living with cystic fibrosis. She wrote of good times and those that were more difficult and recently she wrote of waiting for her transplant. She shared what she did for exercise as well as going through a Nissen Fundoplication surgery. All through her time as a columnist, she was always positive and uplifting with a humor that was distinctly Jennifer.

That she was adventurous and loved life was obvious from reading her quarterly column, "Coughing With A Smile." When she came up with that name, we discussed artwork for it. It was her idea to use a mask and to draw a smile on it because, even though she coughed and at times struggled, she still found humor in life.

While having low lung function, Jennifer wrote of still going to the gym and doing what she could and of trying paddle boarding as a new exercise. When she was younger, Jennifer played softball, tennis and golf – things she had hoped she could once again do after her lung transplant. And she shared with our readers her journey on the road to a lung transplant. How she would wait and be patient. Sadly, she didn't receive the call that would give her new lungs. She died December 5, 2015, at the age of 43.

She shared her spiritual side with quotes that helped get her through the rough patches. But mostly, it was her husband of 18 years, Mark, who really made her life shine. He was her cheerleader when she was not feeling great or was supportive when she had to start thinking of having a lung transplant. She cherished Mark and her good friends for always being there for her. She will be missed by all who knew her.

BOOK REVIEW

A Life In Men

By Gina Frangello

Reviewed by Lisa Cissell

When I was asked to read this book and write a review, I readily said yes. As a 52-year-old woman with CF, who is single and could possibly write my own book about men and dating, I was anxious to see if this book would resonate with me. Little did I know that it would stir many emotions in me and be so much more than a book about dating.

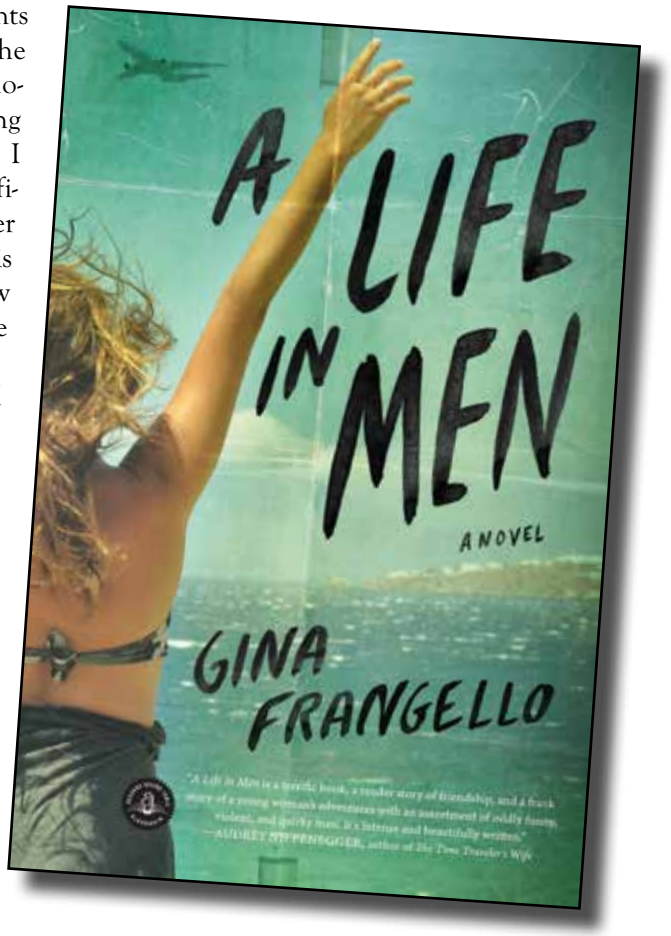
This fictional story tells of the life and travel adventures of Mary, a woman in her 20s who has cystic fibrosis (CF), focusing on the relationships she has with her best friend, Nix, and with the men in her life from boyfriends to long-lost relatives. Although Mary's life starts out sheltered and lukewarm in her suburban life in Ohio, things change dramatically when she decides to join Nix on a trip to Greece. This decision catapults Mary into a totally different life, one filled with love, friendship, adventure, travel and tragedy. Through all of this, Mary tries, sometimes unsuccessfully, to manage her CF and not let it define the woman she is.

Each chapter theme is a different location/man in Mary's life. These chapters are not in chronological order, but move forward and backward in time, so it took me a few chapters to embrace this style. Once I did, it made the book much more enjoyable and I found myself anxious to find where the next chapter would take me. The author is very descriptive in her writing, and I could easily picture the characters, the situations and the locales in my mind. At times, the life experiences portrayed are raw and brutally honest and, for me, that made the story more authentic. Also, it is worth noting that there are several instances of sexual encounters throughout the book, in case that is not your cup of tea.

From the standpoint of a person with CF, I found this book to be factually accurate. There are details of Mary's treatments that include using the Flutter®, taking Pulmozyme® and experiencing episodes of hemoptysis. I felt that the author definitely portrayed a character who was living with this disease and depicted how alone and isolated we sometimes feel.

On a personal level, I could relate to many parts of this book. From Mary having a less common type of genetic mutation that resulted in pancreatic sufficiency, to her love of travel and adventure, this character was like me. The difficulties associated with having relationships and navigating in a "healthy" world were prevalent throughout, but Mary is tough and independent, like so many people I know with CF. A particular scene in which Mary was trying to stifle her cough on a plane took me right back to when I had similar experiences and how conspicuous I felt under the stares of my fellow passengers. But overall, I felt this book was about living your dreams despite your limitations. One phrase in particular struck me, in which Mary was "collecting people and experiences." I took that to mean not collecting as tossing aside, but continuing to connect with people and enjoy life's experiences with them for as long as is humanly possible.

I would recommend this book to adult readers with and without CF. It may be of a bit more interest to women, with relationships and romance being a common theme, but I do think all readers would enjoy the many locations this book takes you, including Greece, London and Kenya. Plus, there are strong characters and a plot that keeps you guessing and wondering what is next for Mary. ▲



Lisa is 52 and has CF. She is a Director of USACFA and is the Secretary. Her contact information is on page 2.

Pay It Forward

BRONZE

Gary & Michelle Allen (in honor of their 20th anniversary and Michelle's 64th birthday)
Derwin & Blanche Ball (in memory of **Marsha Ball-Waldo**)
Rick Birkner
Janice Buck (in memory of **Jimmy Friedeborn**)
Linda Buyaskas (in honor of **Kelly Belly**)
Margaret Clark
Kevin Corr
Nancy Cox (in memory of **C. Barry Woodward**)
Tina Dimick (in memory of **Vincent B. Stoll**)
Pauline DiNello
Maria Domingoes
Bob & Donna Earnest (in memory of **Geoffrey Earnest**)
Andrea Eisenman (in honor of **Beth Sufian & Jim Passamano** for all the good work they do)
Nina Ferrell (in memory of **Amy Young**)
Judy & Ken Greenberg (in memory of **Robert & Leigh Anne Hoehn**)
Becky Hardy
Richard Harris (in memory of **Kathleen Harris** & **John Gary Kevilly**)
Rhonda Hilliard
Abigail Huntington
Carl Keenan
Sara Kominsky (in honor of myself for hanging in there & my 44th birthday)
Jerome F. Lape, Jr. (in honor of his 69th birthday)
Bonnie Larner-Langer
Betty Lefer
Laura Mentch (in honor of **Cathy Chacon, R.N.**)
Jarrod McKee
Prudy Ramsey
Stephanie Rath (in memory of **Jennifer Hale**)
William & Ann Robinson (in honor of **Carl & Kara's** wedding November 22, 2015)
Sheila Schnitzer
Patricia Spadafora (in honor of her 61st birthday)
Jennifer Staashelm
Holly Stewart
Maureen Riordan
Robert & Rochelle Stone (in memory of **Sybil Gertz**)
Penny Stroud
Marilyn Tarr
Alice Todd (in honor of **Cheri Dewilde's** birthday)
David Versteeg (in honor of his 64th birthday)
Jill Walker (in honor of **Jonathan Miller**)
Roger Waring
James Yankaskas

SILVER

Janet King (in memory of son **Thaddeus Novak**)
Laura Tillman (in memory and honor of all those with CF, past and present)

GOLD

Jill Doran

SUSTAINING PARTNER

ModernHEALTH Specialty Pharmacy

DIAMOND SUSTAINING PARTNER

Nancy Wech (in honor of daughter **Lauren Melissa Kelly** and son **Scott W. Kelly**)

TILLMAN continued from page 13

CFTR mutation, and no dose limiting toxicities were observed. The study's primary objective was to evaluate the safety, tolerability and pharmacokinetic profile of N91115. Nivalis Therapeutics is developing a novel class of disease-modifying therapies that are designed to preserve intracellular GSNO (S-nitrosoglutathione), an endogenous molecule with cell-signaling effects that are implicated in the pathophysiology of CF. The company's lead candidate, N91115 initially targets patients with the F508del mutation. Nivalis reports that the trial showed N91115 to be safe and well tolerated over the dose range studied, with the drug's pharmacokinetic profile fully achieved within the targeted blood levels and dose selection rationale.
http://tinyurl.com/qjjekep

Corbus Pharmaceuticals Announces FDA Orphan Drug Designation and Fast Track Status of Resunab™ for the Treatment of CF

Corbus Pharmaceuticals Holdings, Inc., announced that the U.S. Food and Drug Administration (FDA) has designated as a Fast Track development program and granted Orphan Drug Designation to the company's investigational new drug Resunab™ for the treatment of CF. Resunab™ is a novel synthetic oral drug that preferentially binds to the CB2 receptor expressed on activated immune cells and fibroblasts. CB2 activation triggers internal pathways that resolve inflammation and halt fibrosis. Pre-clinical and Phase 1 studies have shown Resunab to have a favorable safety, tolerability and pharmacokinetic profile. It has also demonstrated promising potency in pre-clinical models of inflammation and fibrosis. Resunab triggers the production of "Specialized Pro-resolving Lipid Mediators" that acti-

Continued on page 25

U.S. ADULT CF ASSOCIATION AWARDS

The U.S. Adult CF Association held its first public awards ceremony to present the organization's 2015 Jacoby Angel Award and 2015 Founders Award. The ceremony was held in Phoenix, AZ, on October 9, 2015, during the North American CF Conference and had 102 people in attendance.

Every two years USACFA asks the CF adult community to nominate deserving members in the CF community to be considered for the two awards. After a three-month nomination period, the USACFA Board of Directors votes for the nominee who will receive the award. No Directors can be nominated for an award. If a family member is a nominee then that Director abstains from voting.

The USACFA Founders Award is meant to honor a person who exemplifies the qualities of the founders of USACFA by being an individual who has demonstrated a strong commitment to helping adults with CF. The USACFA founders worked hard to provide a unique newsletter that addressed the needs of adults with CF at a time when there were limited resources for adults with CF. A nominee can be a person with or without CF.

The 2015 Founders Award nominees are an exceptional group. Their experiences span a wide range of areas. Each nominee is a wonderful example of the type of person the award is meant to honor. Nominees included Dr. Michael Boyle, Jessica Martens, Connie St. Clair, Marion Jones and James Passamano. The Founders Award was presented to James Passamano. James is a partner in the law firm of Sufian & Passamano and has spent the past 20 years advocating for the rights of people with CF. James successfully brought a federal civil rights case *pro bono* against Arkansas Medicaid in 2014/2015 regarding the provision of coverage for medically necessary treatment. The case is regarded as one of the most significant victories for the rights of Medicaid beneficiaries in the United States. The case also strengthened the ability of all people with CF to gain access to the treatment they need to fight CF.

The Jacoby Angel Award was established to honor the memory of Dr. Jack Jacoby. Dr. Jacoby was a physician who specialized in the treatment of people with CF. He was a physician at the CF Center at St. Vincent's hospital in New York City. Dr. Jacoby had CF.

Dr. Jacoby worked day and night to try and help his patients at a time when life expectancy was in the teens. He gave everything he could to relieve the suffering of his patients, and he saved the lives of many people with CF. Dr. Jacoby was also the medical advisor for *CF Roundtable* in the 1990s and wrote informative and touching articles for the newsletter. He was and continues to be a hero to many in the CF community. He died in 1997 from liver failure due to CF-related liver disease.

A nominee for the Jacoby Angel Award should be an adult with CF who has made a difference in the lives of one or many people and whose life mirrors the dedication and commitment to helping others that the award's namesake had during his lifetime.

The four Jacoby Angel Award nominees were honored and included Isabel Stenzel Byrnes, Emily Kramer-Golinkoff, William Elder, Jr., and Emily Schiller.

2015 FOUNDERS AWARD NOMINEES JAMES PASSAMANO, MARION JONES, CONNIE ST. CLAIR AND DR. MICHAEL BOYLE ENJOY THE AWARDS CEREMONY.

The Jacoby Angel Award was presented to Isabel Stenzel-Byrnes. Isabel has worked to raise awareness about CF since childhood. She is the co-author of the best-selling book *The Power of Two* and the co-star of the documentary *The Power of Two* portraying the lives of twin sisters with CF and their journey through lung transplant. Isabel was instrumental with her sister, Anabel, in changing a Japanese law that prevented people with CF in Japan from gaining access to life-saving CF medications. Isabel is a longtime volunteer for CFRI and the U.S. Transplant

Games and is a hospice social worker.

The awards ceremony also included a candle lighting ceremony, which honored the memory of Dr. Jack Jacoby, the Founders of USACFA and *CF Roundtable* and for Catherine Martinet and Pammie Post who were founder-like in their contributions to USACFA.

USACFA honored some longtime supporters of the organization and the CF adult community by asking them to be candle lighters. We thank the following for participating in the ceremony: Dr. Pat Walker, Dr. Berdella, Joan Finnegan Brooks, Dr. Marcia Katz, Dr. Noreen Henig, Dr. Ahmet Uluer, Dr. James Yankaskas, Dr. Julie Biller, Dr. Jeff Wine and Marlene Wine, and Paul and Debbie Motenko. The attendees were asked to think about the memory of a person with CF who had inspired them or who inspires them now as the candles were lit.

The awards ceremony included recognition of past Founders Award winners Dr. Jerry Nick and Dr. Yankaskas. Fifteen CF Center social workers were called to the front and presented with small gifts as a token of appreciation for their dedication to helping adults with CF.

The awards ceremony was a memorable event that left many attendees visibly moved by the stories of dedication to the CF community the USACFA awards nominees, recipients and founders exemplify. Many in attendance later remarked the awards ceremony was one of the most memorable events they had attended.

Months after the event, attendees and those who have watched the video continue to express their belief that the awards ceremony was inspiring, moving and an important way to connect with the CF community and honor some outstanding individuals.

USACFA was honored to have many physicians, scientists, nurses, physical therapists, social workers and others who are committed to helping people with CF in attendance. The Adult CF Care Center team from National Jewish in Denver came out in force with 22 members of their team to honor two of their wonderful research nurses. We were proud to be able to recognize so many important contributors to CF medical care, advocacy and research.

To watch the 2015 USACFA awards, go to: http://www.cfroundtable.com/announcements/2015-usacfa-awards/ ▲

USACFA BOARD MEMBERS BETH SUFIAN (LEFT) & MERANDA HONAKER WITH PAST FOUNDERS AWARD RECIPIENTS DR. JERRY NICK (FAR LEFT) & DR. JAMES YANKASKAS.

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.

IN THE SPOTLIGHT

With Reid D'Amico

By Andrea Eisenman and Jeanie Hanley

Some say, "Youth is wasted on the young." They do not know Reid. He wastes no time with all of his accomplishments at the youthful age of 23. After finishing his undergraduate degree at Duke, he is now at Vanderbilt University pursuing his PhD in biomedical engineering. All while volunteering and writing for *CF Roundtable* and *Cystic Fibrosis News Today*.

You may recognize our latest star from previous issues of this newsletter. Reid recently joined our board. He and Meranda Honaker write the Clinical Trials column about the latest medical trials for CF that are in progress as well as what might be in the pipeline soon.

We are fortunate to have him as a Director and a columnist. As you will read from his interview, he brings with him not only his own unique view of having CF but an abundance of knowledge in biomedical engineering and medical knowledge about future possibilities of a cure for CF and other genetic diseases. Please welcome our newest star. Spotlight, please!

Your age at diagnosis: 10

Have siblings? Do they have CF? One brother; he does not have CF.

How was your CF growing up? Did anything healthwise limit you?

I was very fortunate that my CF did not interfere with much growing up. I was able to play sports and be involved in many volunteering and academic activities. Growing up, CF was more of a mental battle. I knew what CF was and what happened to people who had it. I did have a cough growing up, and it made me very self-conscious. Overall, CF was more mental than physical for me.

At what age did you understand what CF was?

Since I was older, I already had an understanding of basic biology and disease. My cousin was diagnosed with CF when she was young, so I was aware of the specifics through my relation to her.

REID D'AMICO

When did you decide you wanted to be a scientist/engineer? Explain with the story of how you decided.

I always had a great love and appreciation for science and engineering. I attended the International Science and Engineering Fair all four years of high school and won awards at the international level. My projects usually centered on renewable energy and the environment. I always had a love for medicine/engineering, but it wasn't until college that I had the opportunity and resources to start to study biomedical engineering.

How did going through college affect your health, if at all?

Duke University is incredibly stressful and rigorous on anyone's body. For me, I had to combine that with my CF. It was difficult to maintain my health (both physical and mental) under the stress, but the help of my friends made it possible. We all went through it together and found solace in each other.

How is your health now and how do you keep it stable to get your PhD?

I'm happy where my health stands. It does not interfere with my studies/personal life. A lot of managing my CF in school came from years of school/life balance I mastered during high school and undergrad. I'm also open about my CF with those who need to know, and they are understanding if I need extra time to finish an assignment.

Explain to lay people what biomedical engineering is.

Biomedical engineering is where medical science and engineering meet. Biomedical engineering is broad and can range from making new medical equipment to manipulating cells to using coding and large amounts of data to understand the human body. It's a powerful new field of study that is advancing the foundation left by basic science.

Is it hard to balance your health with your studies and research?

It used to be, but with time and practice, I've learned to balance. I also love what I do, so any stress that comes with my research is more manageable.

How did you get involved with BioNews Service's *CF News Today*?

During the end of my senior year at Duke, I was taking a light course load and travelling for PhD interviews. I had a lot of down time to read and volunteer. One day, I just decided to send an e-mail and resume to BioNews and explained my back-

ground. They were receptive and gave me my own column to discuss CF and regenerative medicine since it's a field with little work.

What do you do for fun?

I love to hike around Nashville, read, volunteer and write. I usually take advantage of conferences and meetings to explore the city I'm in.

Do you know your mutations for CF?

Homozygous DelF508

Do you see a cure for CF in your lifetime?

Of course! And if not in mine, I hope it won't be long after. We have some amazing scientists and engineers working on manipulating the human genome to rid disease. If we can prove it's safe and effective, we can then work towards finding cures for many genetic diseases.

Who inspires you?

Honestly, I'm inspired by many people. My family and friends who support me despite not always understanding what I do, and my fellow biomedical engineers who spend so many hours trying to cure disease, and the doctors and nurses who keep those with CF alive. I'm surrounded by great people, and I know that all of them have given me inspiration to succeed.

If you could meet with anyone from the past or present, who would it be and why?

I would love to meet my grandfather. He passed away before I was born. He was the only other engineer in my family, and I would love to have that connection with him.

What words of wisdom could you share with others with CF pursuing college?

College is hard for more reasons than just coursework. Find what you love, and set goals—those are what will help you through your toughest weeks. Keep a big picture, and never lose sight of what you want to do.

What do you see yourself doing in five years?

I hope to be close to graduating with my PhD, and I also hope to have more experience with nonprofit volunteering.

Anything else you would want to share with others who have CF?

Use your CF. You understand things that many will not. Use this gift to teach others. ▲

Andrea Eisenman is 51 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. Jeanie Hanley is 53 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.*

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

TILLMAN continued from page 21

vate an internal cascade responsible for the resolution of inflammation and fibrosis. Resunab has direct effects on fibroblasts to halt tissue scarring. In effect, Resunab triggers internal pathways to turn "off" chronic inflammation and fibrotic processes, without causing immunosuppression.

http://tinyurl.com/nthns2b

Study Finds That Pseudomonas Aeruginosa's Ability to Adapt in CF Exacerbates Pulmonary Inflammation

A new study revealed that adaptive changes in Pseudomonas (P.) aeruginosa may exacerbate pulmonary inflammation and contribute to the pathogenesis and progression of chronic lung disease in the context of CF. The majority of CF patients are chronically infected with the opportunistic pathogen P. aeruginosa and have intense neutrophil-associated inflammatory responses that cause lung damage. Since the host immune responses are not effective in eliminating these bacteria, there is persistent bacteria-host interaction, leading to inflammation and immunopathology during CF chronic P. aeruginosa infections. During P. aeruginosa-host interactions, the bacteria genetically adapts to the CF lung milieu. It has been shown that P. aeruginosa isolates are genotypically and phenotypically different from early-stage and environment-based infections. Chronic infection isolates show adaptive changes, such as conversion to mucoidy or loss of motility, reduced expression of acute virulence factors, such as pilus, extracellular toxins and enzymes that cause invasive disease. P. aeruginosa has a transcriptional factor lasR, one of the most important quorum sensing regulators that controls the expression of several exoproducts and acute virulence factors. The authors evaluated the impact of P. aeruginosa lasR mutants on inflammatory responses in vitro, in vivo and in CF patients. They found that lasR mutants induced an exacerbated neutrophil-predominant inflammatory response. Their findings suggest a mechanism by which P. aeruginosa lasR variants from chronic CF infections augment the inflammation of CF lung and potentially increase disease development.

http://tinyurl.com/qxj6tba

Continued on page 29

PARENTING
Support Helps

By Colleen Veitengruber

Hi, my name is Colleen Veitengruber and I was diagnosed with cystic fibrosis (CF) in 1988, at birth. I have been very fortunate and lucky with great health throughout my childhood. I am now 26 years old, a college graduate, a wife, a kindergarten teacher and, finally (and my favorite title), a mother. I have never been in the hospital or been on IV medications, but of course went through my share of rebellious teenage years when I refused to do my treatments. Thankfully, when I was a freshman in college, I met my husband, Tim, who has been completely supportive of me and was the kick-in-the-butt I needed to get my health back on track.

One of my biggest goals in life was to get married and have a family of my own. I have never thought that CF would get in the way of that goal, and after getting married in 2010, Tim and I planned to have children. The road to getting pregnant was a long and bumpy one. It included appointments with genetic counselors, fertility doctors, many tests and procedures, including a minor surgery for me. I had to have one of my fallopian tubes removed because it was damaged from surrounding scar tissue, which was a result of my meconium ileus surgery at birth. After three years, and three IUI (intrauterine insemination) procedures, I was finally pregnant in December of 2013. I was *finally* going to be a mother!

In contrast to the difficult journey of getting pregnant, my pregnancy was relatively easy. Prior to getting pregnant, my lung function was in the 80s and I had also started taking Kalydeco in August of 2013. These factors, in combination with 100 percent compliance to my treatments, I believe, greatly contributed to my easy pregnancy. Throughout my pregnancy, I had many discussions with my fantastic CF doctor about managing my health, taking care of a baby, my interest in nursing her and going back to work as a teacher. My doctor was behind me 100 percent, supporting my decisions to nurse and go back to work. She just kept

COLLEEN VEITENGRUBER AND HER DAUGHTER, ANNA.

reminding me that I needed to keep myself healthy in order to be there for my daughter. I'll admit, I was nervous, but I was so excited and ready to take on the challenge of being a working, nursing CF mommy – nothing was going to hold me back. And on Friday, August 8, 2014, Anna Lily was born weighing in at 6 pounds, 7 ounces at 39 weeks. I was so in love!

After about 48 hours in the hospital, we were sent home – back to reality, a very new reality. I was now a CF mom, and had to learn how to juggle life with a newborn, nursing and keeping on top of my health, with a seven-week maternity leave countdown looming over my head. The first few days/nights were tiring, but I was loving being a mommy. Since I was so excited and always wanting to snuggle our precious little peanut, my treatments often got pushed aside and skipped. Thankfully they weren't neglected for too long before Tim finally made me snap back into reality and reminded me that I needed to take care of myself, too. After Anna's first week home, Tim headed back to work — Anna and I were on our own. I tried to stick to a routine each day so that I wouldn't continue to neglect my treatments, which really helped. Some days were harder than others, forcing myself to set my sweet little baby down next to me while I did my Vest and nebulizers, but it was helpful when the sound of the Vest often lulled Anna to sleep.

After the first month, which flew by, I had to start thinking about going back to work in just a few short weeks. Since I'm a planner and like to know what to expect, when Anna was around five weeks old, she and I did a "trial-run" of a work day. Lucky for us, Tim's Grandma agreed to babysit Anna while both of us are at work. This made me feel so much better knowing that Anna was being taken care of by family and that she would be able to spend time with her cousins every day. So, Tim took Anna to his Grandma's house for the day, while I tried to figure out what my days were going to look like when I went back to work. In the few days prior, I had pumped after nursing Anna so that I could provide her with breast milk

while she was with Grandma. Since Tim would be taking Anna to and from his Grandma's house, this allowed me time to pump and do my treatments before going into work. The first trial run was pretty successful. I stayed back and mostly observed my substitute with my class, learned my daily schedule and figured out when and where I would be able to pump during the school day.

> *"In contrast to the difficult journey of getting pregnant, my pregnancy was relatively easy."*

The following week, Anna and I went to my first post-baby CF clinic appointment — of course my CF doctor was so excited to meet Anna. I was anxious to see how my lung function was holding up and, especially, my weight since I was breastfeeding. I was concerned that my weight was going to be pretty low due to the amount of extra calories needed to provide for Anna. My goal had been to eat almost every time that Anna ate, even if it was just a small snack, anything to get extra calories. I was pleasantly surprised to see my lung function was at my normal baseline with an FEV_1 of 83 percent. I was glad that my weight was also back to my pre-pregnancy weight and not lower. My CF doctor encouraged me to keep nursing since it was going so well and to keep on top of my treatments so that I can be as healthy as possible for my daughter.

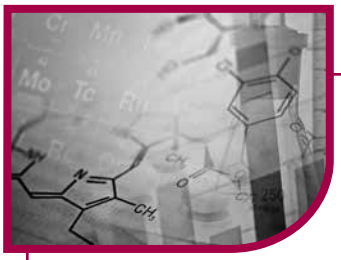
After a successful second trial run on the Friday before my first day back, it was finally time for me to become an official working mommy. The first weeks were definitely an adjustment. It was hard leaving my child and going to take care of and teach twenty-four others. I had to remind myself *"this is normal; there are plenty of women who go back to work and whose children are in daycare; I'm doing what's best for Anna by working and helping to support our family."* It was also an adjustment realizing how much less time I would be spending with Anna during the week, but my favorite times of each day soon became nursing her as soon as she got home and first thing in the morning. This was our time to bond and I was so proud of my "sick" body for being able to provide for her.

I am very lucky to be able to exclusively nurse my daughter for four months now. It has not been easy, especially being back at work, but my co-workers have been very supportive of my decision. I have a wonderful teaching assistant who takes my kids out for recess every day so that I can have a pumping break (and do my Cayston if it's an "on" month). Also, our Instructional Coach has kindly blocked a half hour each Tuesday afternoon, when I don't have a plan time, to supervise my class and allow me to use her office, so I can pump. I typically pump in my classroom right before school, during recess and during my afternoon plan time. If for whatever reason I am unable to use my classroom (ex: indoor recess), I have used our Conference Room, the Social Worker's office, and the Instructional Coach's office. It's wonderful, and very helpful, knowing I have the support of my co-workers to help me provide the best I can for my child.

Since Tim was taking Anna to and from Grandma's house, this meant that there are about two hours between the time my students go home and when Anna (and Tim) would be home. For the first few weeks, I used this time to get caught up on schoolwork and plan for the upcoming days and weeks. I felt very behind and out of the loop since I didn't start the school year with my class, so this time was used to catch up and get back into the swing of things. I always made sure that I was home as soon as Anna got home so I could nurse her right away. Since I was staying after school for an hour or two until Anna and Tim came home, this meant I would have to do my evening treatment after feeding Anna. At first this really bugged me because it was taking away a bit of the few hours I get with Anna before we both would crash for the night. But I reminded myself that even though I can't spend those thirty minutes with her, I know that it's better for us long-term, and that my treatments are allowing me to spend more months and years as her mother. Now that I have been back to work for a couple months, I am able to plan while pumping and I only stay after school for an hour or so, and I try to get home before Anna and Tim so I can finish my treatments before they arrive.

Being a working, nursing CF mommy has been both challenging and so rewarding at the same time. I am extremely proud of my body for carrying and delivering a beautiful, healthy little girl and for continuing to provide all of the nutrients that she needs. It's amazing, even to me, to defy the odds of CF and to be "beating" cystic fibrosis every day. I am so thankful for Tim, our family, friends and co-workers for their continued encouragement and support of this amazing journey called parenthood. ▲

Colleen is 26 and has CF. She and her husband, Tim, live in Decatur, IL, with their daughter, Anna, who was successfully breastfed until 13 months of age. Colleen now is teaching third grade. She loves to connect with others who have CF and their families. You may contact her at: colleen.veitengruber@gmail.com.



SEARCHING FOR THE CURE

Sinus Clinical Trials: Cystic Fibrosis And Sinus Complications

By Reid D'Amico

Sinus complications tend to go forgotten when it comes to cystic fibrosis (CF). Years ago, mortality surrounded the malabsorption as a result of pancreatic insufficiency. Today, the majority of cystic fibrosis complications concern the lungs. In CF, the epithelial cells in the lungs produce mucus that is abnormally thick. This tenacious mucus is difficult to clear from the lungs, and bacteria become trapped and result in infection. Physiologically, the lung epithelial cells and the nasal and sinus epithelial cells are similar, so it is no surprise that CF sinuses tend to fall victim to infections as well. Sinus infections cause the sinuses to become inflamed. This inflammation, like inflammation in the lung, causes damage. This chronic infection and inflammation in the sinuses can lead to the development of nasal polyps that can further obstruct the sinuses and lead to more infection. The polyps are cysts that can form anywhere in the nasal passage and sinuses. Due to their obstructive nature, the polyps further prevent the thick sinus mucus from draining.

So why are sinus complications rarely discussed or known by others outside of CF? While there is no correct answer, it likely has to do with the anatomical and physiological differences between the lung and the sinuses. Infection and damage to the lungs interferes with the body's ability to exchange carbon dioxide and oxygen. Without this gas exchange, the body and its tissues are starved of oxygen, which ultimately results in death. When it comes to the sinuses, there is no gas exchange or physiological occurrence that is absolutely necessary to sustain human life. While the sinuses and nasal passage are an important respiratory and sensory organ, their infection and damage is largely seen as a limitation to one's quality of life. However, this view of the sinuses is not sustainable with the life expectancy increasing for those with CF. A compromised nasal passage and sinus cavity can lead to crippling headaches along with other complications that can impair someone's life.

Some research has been done regarding drug delivery to the sinuses. From personal experience, many with CF know that antibiotics can be hit or miss—with some antibiotics not adequately addressing infection and some working to knock down infection. For some, this issue tends to become more relevant in the sinuses. In fact, scientists and engineers have drifted away from the conventional approach of blood-delivered medications and are now researching medication that can be delivered to the sinuses directly. Biomedical engineers are working on creating biomaterials that can be inserted into the nasal passage to deliver a therapeutic benefit. Some devices have passed clinical trial and are being used to reduce the inflammation seen in chronic sinusitis. The issue for people who have CF, however, is that inflammation is coupled as a secondary effect to infection. There is a great need to develop biomaterials that can be implanted into the nose or sinus that can deliver antibiotics. By directly delivering antibiotics to the infected sinus and nasal passage, we can hope that many of the challenges that affect the quality of life for those with cystic fibrosis can be addressed.

> *"Sinus infections cause the sinuses to become inflamed. This inflammation, like inflammation in the lung, causes damage."*

REID D'AMICO

Reid is 23 and has CF. He is a Director of USACFA. His contact information is on page 2.

December 2015 Clinical Trial Spotlight

1. Anthera: Solution Study of Oral Liprotamase Unit-Matched Therapy of Non-Porcine Origin in Patients with Cystic Fibrosis
https://clinicaltrials.gov/show/NCT02279498

2. ProQR: Dose Escalation Study of QR-010 in Homozygous DeltaF508 Cystic Fibrosis Patients
https://clinicaltrials.gov/show/NCT02532764

3. ProQR: Exploratory Study to Evaluate QR-010 in Cystic Fibrosis Subjects Homozygous and Heterozygous DeltaF508 Mutation
https://clinicaltrials.gov/show/NCT02564354

4. Nivalis: Study of N91115 in Patients Homozygous for the F508del-CFTR Mutation (SNO-6)
https://clinicaltrials.gov/show/NCT02589236

5. Role of Glucagon in Glucose Control in Cystic Fibrosis Related Diabetes
https://clinicaltrials.gov/show/NCT02398383 ▲

TILLMAN continued from page 25

New guidelines for Pseudomonas aeruginosa eradication

The U.S. CF Foundation has published new guidelines for the eradication of Pseudomonas (P.) aeruginosa. The three recommendations are:

1. Inhaled antibiotic therapy is recommended to treat first-growth P. aeruginosa. The preferred regimen is tobramycin inhaled solution (TIS) 300 mg BID. for 28 days. A longer treatment course or the addition of oral ciprofloxacin is not recommended.

If the initial eradication course fails, the committee stated that the use of additional or other antibiotics (inhaled, oral or IV) would be a reasonable first step.

2. The use of prophylactic antipseudomonal antibiotics is not recommended. The EPIC trial showed that routine prophylaxis provided no benefit over initiating treatment at first isolate. The committee also noted that prolonged use of antipseudomonal antibiotics may increase the risk of acquiring other pathogens.

3. For patients who cannot expectorate sputum, routine oropharyngeal (OP) cultures are recommended to detect P. aeruginosa infection. Airway cultures should be obtained every three months. A recent review reported that there was no evidence to support the use of routine bronchoalveolar lavage rather than relying on OP cultures and clinical symptoms. The committee noted that a more aggressive approach to eradication may be warranted but further study is required.
http://tinyurl.com/odw3zya

Honey for Burkholderia cepacia?

Manuka honey has acquired a reputation as an alternative therapy, and a new study suggests that it may be a useful add-on therapy for Burkholderia and P. aeruginosa infections. Testing was performed on 111 strains of Burkholderia and P. aeruginosa to determine their susceptibility to manuka honey, tobramycin and colistin. All strains demonstrated susceptibility to manuka honey. MICs for four representative strains showed additive or synergistic activity when honey was combined with an antibiotic. The authors concluded that honey may be a useful agent in combination with antibiotics to lower the MIC needed to inhibit Burkholderia and P. aeruginosa, but further study is needed.
http://tinyurl.com/zmcyzeg

U of T research sheds new light on mysterious fungus that has major health consequences

Researchers at the University of Toronto examined fungi in the mucus of patients with CF and discovered how one particularly cunning fungal species has evolved to defend itself against neighboring bacteria. A regular resident of our microbiome — and especially present in the lungs of CF patients — the Candida albicans fungus is an "opportunistic pathogen." Candida albicans is a particularly wily fungus. Its signature maneuver is shapeshifting — it can morph from a round, single-celled yeast into a long stringy structure, allowing it to adapt to different environments and making it exceptionally harmful. As part of an ongoing battle between microbes, certain bacteria, which are also found in CF patients, secrete molecules preventing the fungus from changing into its stringy shape. The researchers tried exposing the mutated fungus to these bacterial rivals. Instead of responding to the bacterial signals, the fungus kept to its stringy form. The researchers believe these fungi have evolved to counter the tactics of their bacterial rivals.
http://tinyurl.com/qb38svq

OrPro Therapeutics Receives U.S. Patent Covering Novel Enzymatic Treatment for CF

OrPro Therapeutics, Inc., announced today that the U.S. Patent and Trademark Office has issued a U.S.

Continued on page 30

TILLMAN continued from page 29

Patent covering the ability of Theradux™ (recombinant modified human thioredoxin-1) to improve the viscoelastic properties of mucus secretions in patients with CF. Theradux is an inhaled, non-systemic version of the naturally-secreted enzyme thioredoxin, which helps to prevent excess molecular bonds from forming within the mucus layer. Treatment with nebulized Theradux, which relaxes abnormally stiffened mucus, is anticipated to help restore mucus transport and clearance in the majority of CF patients. Theradux is expected to work both alone and in concert with approved and in-development CF Transmembrane Regulator (CFTR) modifier.
http://tinyurl.com/hh95puu

FDA approves digestive enzyme cartridge for patients on enteral feeding

Alcresta Pharmaceuticals has announced it received de novo approval from the U.S. Food and Drug Administration (FDA) to market Relizorb™ (immobilized lipase). Relizorb is a first-of-its-kind digestive enzyme cartridge designed to mimic the normal pancreatic function by breaking down fats in enteral tube feeding formula. By breaking down these fats from enteral tube feeding formulas prior to ingestion, Relizorb allows for the delivery of increased absorbable calories from fatty acids and monoglycerides to adults who are partially or completely unable to break down and absorb fats. Relizorb is developed using Alcresta's proprietary enzyme immobilization technology. The active ingredient in Relizorb is the digestive enzyme lipase, attached to polymeric carriers together called iLipase®. As the enteral tube feeding formula passes through Relizorb, it makes contact with the iLipase and the fat in the formula is broken down to its absorbable form (fatty acids and monoglycerides) prior to ingestion. The iLipase remains in the cartridge and does not become part of what is ingested.
http://tinyurl.com/z4gc36a

Synedgen Presents New CF Complication Treatment Options at North American CF Conference

Researchers at Synedgen are developing targeted therapies that address complications of CF on pulmonary and GI surfaces. Synedgen is harnessing the science of glycomics to discover and develop a new class of polysaccharide-based drugs that targets the complex interactions at the cell surface and mucosal interface. What Synedgen calls its "Glycomics Technology Platform" enables its scientists to design drugs designed to suppress inflammation, reduce infection and improve healing, and leverage this unique, proprietary technology engine to create new molecular entities that control the desired cellular response and direct activity at the source of the selected disease. Synedgen says its chemistry advantage has resulted in development of a scientifically validated pipeline of drugs able to:

- Suppress inflammation and improve healing.
- Loosen mucus and biofilms to enhance their elimination.
- Reduce mucus and biofilm adhesion to mucosal surfaces.
- Reduce the ability of bacteria to bind and invade tissues.
- Protect mucosal surfaces from damage and microbial imbalances.

http://tinyurl.com/pkegj65

Galapagos and Abbvie Present Encouraging CF Discovery at #NACFC

The novel potentiator GLPG1837, a drug candidate to treat the Class III CF mutation, has demonstrated that it is safe and well-tolerated in a randomized, double-blind, placebo-controlled Phase I study conducted over a range of single and multiple doses in healthy human volunteers in Belgium. The research also revealed favorable drug-like properties in GLPG1837 as well as no significant adverse events. Galapagos announced during the conference that the encouraging results support the initiation of its Phase II study in Class III mutation patients, which is planned to begin at the end of the year. In addition, the company expects to start studying GLPG1837 in combination with other Galapagos candidate drugs to evaluate its use as a potential triple combination treatment for people with Class II CF, the mutation group with the largest amount of patients. Galapagos and Abbvie are working together following the signing of a collaboration agreement focused on the development of novel CF treatments. While the Phase 1 trial for the GLPG1837 potentiator has been completed, a new Phase 1 study for the GLPG2222 corrector is expected to be initiated by the end of the year.
http://tinyurl.com/qanhfzs

Proteostasis Therapeutics, Inc., Demonstrates Therapeutic Potential of a Proprietary Triple Combination Therapy for the Treatment of CF That Restores Activity of Mutant F508del CFTR Protein to 80% of Normal

Proteostasis Therapeutics, Inc. (PTI), announced the expansion of its CF drug candidate pipeline to include the addition of a novel triple combination therapy of PTI's own CFTR amplifiers, correctors and potentiators. PTI's testing has shown that a triple combination comprising a proprietary corrector and potentiator and one of PTI's CFTR amplifiers, can restore the activity of mutant F508del CFTR protein to 80% of normal activity. PTI is advancing its CFTR amplifier PTI-428 as its lead clinical development candidate for the treatment of CF and expected to file an IND with the FDA by the end of 2015. The PTI correctors and potentiators are expected to enter clinical development by the middle of 2017.
http://tinyurl.com/nptup6r

Microscopic view of coughed-up mucus may be new biomarker for CF progression

A team of researchers at the Center for Nanomedicine at the Wilmer Eye Institute,

part of Johns Hopkins University School of Medicine, has been studying mucus in the lungs of CF patients. Their primary goal was to design inhalable therapeutic nanoparticles that cross the CF mucus barrier in the lung, to help restore normal function. But the work recently led the researchers to the unexpected discovery that mucus appears to change as the disease progresses. They found that the mobility of these nanoparticles could vary widely in mucus from different patients. They explored these differences by using a technique called "multiple particle tracking," which allowed them to take videos of nanoparticle movement within CF mucus, using a fluorescent microscope. In terms of applications for their findings, there are currently no available biomarkers that reliably predict pulmonary exacerbations, a drastic decline in lung health. That knowledge would help to better manage CF patient care. The hope is that this approach will provide more insights into the CF lung microenvironment that are important to maintaining proper lung function.
http://tinyurl.com/zn7fvw7

Scripps scientists find protein flaw in CF

The CF transmembrane conductance regulator protein is so busy communicating with the wrong neighboring cells that it can't function normally and is prematurely degraded. By removing the chatter, researchers partially restored the protein's normal function. The findings suggest that therapies could one day treat the root cause of CF, not just the symptoms. When the researchers analyzed cell samples, they identified almost every protein CFTR interacted with — even tracking the secondary and tertiary protein interactions. While it was thought that most mutant proteins just lack one or two crucial interactions, the CFTR mutant had acquired an entirely new network. The researchers narrowed these mutant protein interactions to just eight key disruptive proteins, used a gene silencing approach to block the interaction of these proteins with the mutant CFTR, and found that the CFTR protein partially returned to normal function.
http://tinyurl.com/zkd6m5h
and
http://tinyurl.com/ovcpslo

Antibiotic Resistance in CF Patients Explored Using Whole Genome Sequencing

Authors profiled 28 bacterial strains (that are not Pseudomonas aeruginosa) isolated from three children with CF using whole-genome sequencing to detect if indeed there were indicators for antibiotic resistance or unique virulence traits. The team detected that two bacteria strains – Staphylococcus aureus and Achromobacter xylosoxidans – were resistant even in the presence of appropriate antibiotic therapy, which suggests that the antibiotic treatment may not reach susceptible populations of bacteria. Possible mechanisms responsible for this phenotype are the existence of physical barriers and inactivation of antibiotics action. The multi-drug resistant Achromobacter xylosoxidans was capable of persisting for almost five years, while the same was not observed for another isolated multi-drug resistant strain, the Steno-trophomonas maltophilia.

Altogether, the authors noted that their findings identified non-pseudomonal bacteria species in CF patients and particular attention should be given to Achromobacter xylosoxidans, since it can persist for long periods of time, possibly by mediating antibody resistance.
http://tinyurl.com/j9lbm7c

Anti-smoking drug Inv102 could help treat asthma, CF

Inv102 works by blocking a signaling process called the beta arrestin pathway that tells the cells lining a person's lungs to transform and begin producing mucus when they are impacted by smoke or disease — preventing lungs from becom-

Continued on page 32

The CFLF Is In Search Of Board Members!

The Cystic Fibrosis Lifestyle Foundation (CFLF) is about guiding the choices made to live successfully with CF.

Through Recreation Grant awards, the CFLF assists people living with CF to thrive, not just survive. By inspiring healthy and active lifestyles through fitness, exercise, and recreation activities the CFLF educates people living with cystic fibrosis about the critical psychological, social and emotional connections between their lifestyle and their health.

To date, the CFLF has awarded more than 750 grants totaling more than $420,000 in direct assistance, but the demand for help continues to increase. The organization needs your help in meeting this growth.

For more details on new Board Member positions go to cflf.org/board-member-description

To submit an application for becoming a CFLF Board of Directors candidate, please send a one-page letter of interest and current résumé or CV to:

boardapplication@cflf.org

For more information or questions, please contact:
Brian Callanan - brian@cflf.org

CFLF
Cystic Fibrosis Lifestyle Foundation

ing clogged with mucus, causes a person to cough or have difficulty breathing. It does this by binding a molecule onto the beta receptors on the surface of the lung so they cannot send out the signal to transform into mucus-producing cells. As the blocked cells die they are replaced by regular epithelial cells, rather than mucus-producing cells, effectively breaking the cycle. While the anti-smoking version of the drug is a pill, Dr. Collier said an inhaled version that directly coats the lungs of people with asthma, CF and other severe respiratory conditions was being developed.
http://tinyurl.com/oqk2tw4

Raptor Pharmaceutical Completes Acquisition of Quinsair™

Raptor Pharmaceutical Corp. announced that it has completed the acquisition of Quinsair™. Quinsair is a proprietary inhaled formulation of levofloxacin, a fluoroquinolone antibiotic, which is approved in the European Union and in Canada for the management of chronic pulmonary infections due to Pseudomonas aeruginosa in adult patients with CF. Administration of Quinsair with a high efficiency eFlow Nebulizer System allows for the delivery of high concentrations of active drug directly to the site of infection in approximately five minutes. Quinsair is contraindicated in patients with hypersensitivity to levofloxacin, a history of tendon disorders related to fluoroquinolones, epilepsy or who may be pregnant or breast feeding. Quinsair's safety was evaluated in two double-blind, placebo-controlled studies and in an active comparator study in which the most frequently reported adverse reactions were cough/productive cough, distortion of sense of taste and fatigue/asthenia.
http://tinyurl.com/h8tkz54

PATHOGENS

Comparing the harmful effects of nontuberculous mycobacteria and gram negative bacteria on lung function in patients with CF. Qvist T, Taylor-Robinson D, Waldmann E, Olesen HV, Hansen CR, Mathiesen IH, Høiby N, Katzenstein TL, Smyth RL, Diggle PJ, Pressler T. J Cyst Fibros. 2015 Oct 5

To better understand the relative effects of infection with nontuberculous mycobacteria and gram negative bacteria on lung function decline in CF, the authors assessed the impact of each infection in a Danish setting. They demonstrated the impact on lung function of each chronic CF pathogen. M. abscessus complex was associated with the worst impact on lung function. Eradication of M. abscesses complex may significantly improve lung function.
http://tinyurl.com/o3x4r87

Prevalence of Mycobacterium lentiflavum in CF patients, France. Phelippeau M, Dubus JC, Reynaud-Gaubert M, Gomez C, Stremler le Bel N, Bedotto M, Prudent E, Drancourt M. BMC Pulm Med. 2015 Oct 26;15:131

M. lentiflavum was the third most common mycobacteria isolated in CF patients, particularly in six male patients. M. lentiflavum outbreaks are emerging in CF patients.
http://tinyurl.com/q25jty7

A longitudinal analysis of chronic MRSA and Pseudomonas aeruginosa co-infection in CF: A single-center study. Maret L. Maliniak, Arlene A. Stecenko, Nael A. McCarty. Journal of CF. Published Online: November 20, 2015

Data from the Atlanta Care Center suggest that chronic MRSA and PA co-infection may be associated with increased rate of lung function decline and rate of intravenous antibiotics compared with patients with either pathogen alone.
http://tinyurl.com/qzoka2l

TREATMENTS/INFECTION CONTROL

Subinhibitory Doses of Aminoglycoside Antibiotics Induce Changes in the Phenotype of Mycobacterium abscessus. Tsai SH, Lai HC, Hu ST. Antimicrob Agents Chemother. 2015 Oct; 59(10):6161-9

In this study, the effects of subinhibitory doses of aminoglycoside antibiotics on Mycobacterium abscessus were investigated. The treatment of M. abscessus cells with subinhibitory doses of amikacin was found to change their colony from a smooth to a rough morphotype and increase their ability to adhere to a polyvinylchloride plate, aggregate in culture, and resist phagocytosis and killing by macrophages. M. abscessus cells treated with a subinhibitory dose of amikacin also became more potent in Toll-like receptor 2 (TLR-2) stimulation, leading to increased tumor necrosis factor alpha (TNF-) production by macrophages. These findings suggest the importance of using sufficient doses of antibiotics for the treatment of M. abscessus infections.
http://tinyurl.com/ht8mnx2

Infection control knowledge, beliefs and behaviors amongst CF patients with epidemic Pseudomonas aeruginosa. Somayaji R, Waddell B, Workentine ML, Surette MG, Brager NP, Rabin HR, Parkins MD. BMC Pulm Med. 2015 Nov 5;15:138

Epidemic P. aeruginosa (ePA) infections are common in CF and have been associated with accelerated clinical decline. Factors associated with ePA are unclear, and evidence-based infection control interventions are lacking. The authors found that infections with ePA are closely linked to past exposures, now routinely discouraged. As socialization is the greatest risk factor for ePA, infection control strategies for ePA must focus on discouraging face-to-face interactions among CF patients. As peer support remains a desire amongst patients, investment in technologies and strategies that enable indirect communication and support are required.

http://tinyurl.com/jma699q

Prolongation of antibiotic treatment for CF pulmonary exacerbations. Valerie Waters, Sanja Stanojevic, Michelle Klinger, Jackie Chiang, Nicole Sonneveld, Richa Kukkar, Elizabeth Tullis, Felix Ratjen. Journal of CF. November 2015. Volume 14, Issue 6, Pages 770–776

Pulmonary exacerbations frequently lead to an irrevocable loss of lung function in CF patients. Although extended antibiotic duration has not been shown to be associated with improved outcomes in CF overall, it is not known whether there is a subset of patients who may benefit from longer treatment courses.

This study highlights that in the treatment of pulmonary exacerbations, maximum lung function is not achieved within 14 days in all patients, and that there is continued improvement beyond this period.
http://tinyurl.com/ngqjtum

Factors associated with response to treatment of pulmonary exacerbations in CF patients. Valerie J. Waters, Sanja Stanojevic, Nicole Sonneveld, Michelle Klingel, Hartmut Grasemann, Yvonne C.W. Yau, Elizabeth Tullis, Pearce Wilcox, Andreas Freitas, Mark Chilvers, Felix A. Ratjen. Journal of CF. November 2015.Volume 14, Issue 6, Pages 755–762

Pulmonary exacerbations are associated with significant lung function decline from baseline in CF, and it is not well understood why some patients do not respond to antibiotic therapy. The objective of this study was to identify factors associated with lung function response to antibiotic treatment of pulmonary exacerbations. The authors conclude that inadequate reduction of inflammation during an exacerbation is associated with failure to recover lung function and increased risk of subsequent re-exacerbation in CF patients.
http://tinyurl.com/nvtrql8

Randomized trial of efficacy and safety of dornase alfa delivered by eRapid nebulizer in CF patients. Gregory S. Sawicki, Will Chou, Karina Raimundo, Ben Trzaskoma, Michael W. Kinston. Journal of CF. November 2015. Volume 14, Issue 6, Pages 777–783

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. The authors conclude that administration of dornase alfa via the eRapid nebulizer resulted in comparable efficacy and safety, shorter nebulization times and higher patient preference.
http://tinyurl.com/jaggu55

Antimicrobial Activity of Fosfomycin-Tobramycin Combination against Pseudomonas Aeruginosa Isolates Assessed by Time-Kill Assays and Mutant Prevention Concentrations. Díez-Aguilar M, Morosini MI, Tedim AP, Rodríguez I, Aktaş Z, Cantón R. Antimicrob Agents Chemother. 2015 Oct;59(10):6039-45

The antibacterial activity of fosfomycin-tobramycin combination was studied by time-kill assay in eight P. aeruginosa clinical isolates belonging to the fosfomycin wild-type population but with different tobramycin susceptibilities. The results suggest that fosfomycin-tobramycin can be an alternative for infections due to P. aeruginosa since it has demonstrated synergistic and bactericidal activity in susceptible isolates and those with low-level tobramycin resistance. It also prevents the emergence of resistant mutants in either aerobic or anaerobic environments.
http://tinyurl.com/q5baaoj

FYI

Pilot trial of light therapy for depression in hospitalized patients with CF. Kopp BT, Hayes D Jr, Ghera P, Patel A, Kirkby S, Kowatch RA, Splaingard M. J Affect Disord. 2016 Jan 1;189:164-8.

Depression is common in CF and linked with worse outcomes during hospitalization. Bright-light therapy during hospitalizations augments antidepressant regimens and reduces length of stay in depressed non-CF patients, but has not been examined in CF. The authors found that light therapy was well tolerated by hospitalized CF patients and resulted in improved depressive symptoms and quality of life. Light therapy was associated with a reduced length of stay. Large, randomized trials of light therapy may be indicated for hospitalized CF patients.
http://tinyurl.com/p74vmfh

International Committee on Mental Health in CF: CF Foundation and European CF Society consensus statements for screening and treating depression and anxiety. Thorax 10.1136/thoraxjnl-2015-207488

Studies measuring psychological distress in individuals with CF have found high rates of both depression and anxiety. Psychological symptoms in both individuals with CF and parent caregivers have been associated with decreased lung function, lower body mass index, worse adherence, worse health-related quality of life, more frequent hospitalizations and increased healthcare costs. To identify and treat depression and anxiety in CF, the CF Foundation and the European CF Society invited a panel of experts, including physicians, psychologists, psychiatrists, nurses, social workers, a pharmacist, parents and an individual with CF, to develop consensus recommendations for clinical care. Fifteen guideline recommendation statements for screening and treatment of depression and anxiety in individuals with CF and parent caregivers were finalized by vote. As these recommendations are implemented in CF centers internation-

Continued on page 34

TILLMAN continued from page 33

ally, the process of dissemination, implementation and resource provision should be closely monitored to assess barriers and concerns, validity and use.
http://tinyurl.com/ouogpex

Glucose intolerance in CF as a determinant of pulmonary function and clinical status. Moran Lavie, Dor Fisher, Daphna Vilozni, Rinat Forschmidt, Ifat Sarouk, Hannah Kanety, Rina Hemi, Ori Efrati, Dalit Modan-Moses. Diabetes Research and Clinical Practice. December 2015. Volume 110, Issue 3, Pages 276–284

CF-related diabetes (CFRD) is associated with a decrease in pulmonary function and nutritional status. The clinical significance of impaired glucose tolerance (IGT) in CF patients was investigated. It was determined that IGT in CF patients is associated with increased inflammation and decreased nutritional status and pulmonary function.
http://tinyurl.com/ztr4brh

Imaging modalities in CF: emerging role of MRI. Wielpütz MO, Mall MA. Curr Opin Pulm Med. 2015 Nov;21(6):609-16

MRI is sensitive enough to detect hallmarks of CF lung disease such as bronchial wall thickening, bronchiectasis, mucus plugging and abnormal lung perfusion. A morpho-functional MRI score has been established for semiquantitative assessment of these characteristic abnormalities over a broad range of disease severity. Recent studies demonstrated that MRI is sensitive to detect changes in mucus plugging and lung perfusion in response to antibiotic therapy for pulmonary exacerbations. These results suggest that MRI may be suitable for noninvasive monitoring and as a quantitative endpoint in clinical trials for CF.
http://tinyurl.com/qejhup2

Forced Expiratory Volume in 1 Second Variability Helps Identify Patients with CF at Risk of Greater Loss of Lung Function. Morgan WJ, VanDevanter DR, Pasta DJ, Foreman AJ, Wagener JS, Konstan MW; Scientific Advisory Group and the Investigators and Coordinators of the Epidemiologic Study of CF. J Pediatr. 2015 Sep 18

This study was designed to evaluate several alternative measures of forced expiratory volume in 1 second percent predicted (FEV_1 %pred) variability as potential predictors of future FEV_1 %pred decline in patients with CF. The authors found that median deviation from the best FEV_1 %pred is a simple metric that markedly improves prediction of FEV_1 %pred decline even after the inclusion of demographic and clinical characteristics and the FEV_1 %pred rate of decline. The routine calculation of this variability measure could allow clinicians to better identify patients at risk and, consequently, in need of increased intervention.
http://tinyurl.com/zlaxtxn

Renin-associated hypertension after bronchial artery embolization in CF. Nathalie Coolen, Hervé Gouya, Reem Kanaan, Isabelle Honoré, Jeanne Chapron, Dominique Hubert, Paul Legmann, Daniel Dusser, Pierre-Régis Burgel. Journal of CF. Published Online: September 24, 2015

Bronchial artery embolization is the recommended therapy for massive hemoptysis in patients with CF. This study reports on two cases of multiple renal infarcts and renin-associated hypertension and hypokalemia occurring in CF adults after bronchial artery embolizations. These complications were presumably related to crossing of small calibrated microspheres through arteriovenous anastomoses. Although hypokalemia resolved rapidly, hypertension persisted at least six months and its control required multiple antihypertensive agents. Physicians should be aware of this potentially severe, but previously unreported, complication of bronchial artery embolization.
http://tinyurl.com/hc6yh97

Web-based symptom screening in CF patients: A feasibility study. Balzano J, Fresenius A, Walker P, Berdella M, Portenoy RK, Bookbinder M, Glajchen M, Plachta A, Langfelder-Schwind E, Chen J, Dhingra L. J Cyst Fibros. 2015 Nov 20

CF causes high illness burden. Screening may identify patients who could potentially benefit from interventions for symptoms or other sources of distress. The authors evaluated the feasibility of a web-based system for routine monitoring and found that repeated web-based screening for symptom distress and advance care planning preferences is feasible in adult CF patients. Future studies should assess the system's generalizability and staff resources when implementing reminders and non-web methods of completion.
http://tinyurl.com/zty4ecn

New developments in inhaler devices within pharmaceutical companies: A systematic review of the impact on clinical outcomes and patient preferences. Ninane V, Vandevoorde J, Cataldo D, Derom E, Liistro G, Munghen E, Peché R, Schlesser M, Verleden G, Vincken W. Respir Med. 2015 Nov;109(11):1430-8

The aim of this research was to evaluate how far new developments of inhaler devices are scientifically supported and translate into improvements of patient preferences and/or clinical outcomes. A total of 30 studies were found comparing Respimat® vs. HandiHaler®, Diskus®/Accuhaler® vs. Diskhaler®/Rotadisk® or pMDI, Ellipta® vs. Diskus®/Accuhaler®, Nexthaler® vs. pMDI, or Breezhaler® vs. Aerolizer®. These studies show that developments of inhaler devices may improve patient satisfaction but do not lead to demonstrable improvements in clinical efficacy. Current changes of devices are most commonly paralleled by changes in administration frequency towards once-daily treatment. The only well-documented effect was found for the Respimat® Soft

CF ROUNDTABLE SUBSCRIPTION FORM (Please Print Clearly)

PLEASE CHECK ONE: ☐ **NEW SUBSCRIPTION** **OR** ☐ **UPDATE**

NAME _____ **PHONE (** **)** _____

ADDRESS _____

CITY _____ **STATE** _____ **ZIP+4** _____ **+** _____

E-mail address: _____

I am interested in CF Roundtable because I am a: CF ADULT _____ BIRTHDATE _____

OR I am a: Parent ____ Relative ____ Friend ____ Medical/Research Professional ____ Caregiver ____

CF Roundtable is available at no cost. However, to help defray printing and mailing costs, donations are always welcome.

- Donation to defray costs . . . . . $ _____
- Please send me the back issues I have listed on a separate sheet of paper.
- Please send subscriptions to the names and addresses I have listed on a separate sheet of paper.

Please do not write in this space

WI 16

For donations, make checks payable to USACFA and mail with this completed form to:
USACFA, P.O. Box 68105, Indianapolis, IN 46268-0105
We ask that everyone keep their home and e-mail address information up to date.

OR

You can also subscribe online! Go to www.cfroundtable.com. You can now scan this QR Code with your smart phone to go directly to our online registration page. By subscribing online you have the ability to download a PDF of the latest newsletter, receive the mailed version or you can receive both. The online version will be available two weeks prior to the mailed version.

KEEPING YOUR INFORMATION CURRENT

To keep our records up to date, please be sure to complete and return a subscription form, on this page, to us or register online with any changes to your information, www.cfroundtable.com. (Any issue of the newsletter that is returned to us costs us about two-and-a-half times the first-class postage rate for that piece. Currently that runs about $3.73 per returned copy.) **Thank you for helping us with this.**

Mist™ Inhaler, which realizes a more than three-fold lowering of the once-daily tiotropium dose through increased performance of the inhaler device. There are however, no data on clinical efficacy or safety comparing the two devices at the same dosage. http://tinyurl.com/jlk6p82

TRANSPLANTATION

Epidemic Pseudomonas aeruginosa infection in patients with CF is not a risk factor for poor clinical outcomes following lung transplantation. Julia Pritchard, Mitesh V. Thakrar, Ranjani Somayaji, Michael G. Surette, Harvey R. Rabin, Doug Helmersen, Dale Lien, Swathi Purighalla, Barbara Waddell, Michael D. Parkins. Journal of CF. Published Online: December 02, 201

Unlike pre-transplant outcomes, CF patients infected with epidemic Pseudomonas Aeruginosa (ePA) do not experience worse post-transplant outcomes than those infected with unique strains. Therefore, lung transplantation should be considered for all patients with P. aeruginosa infection and end stage lung disease, irrespective of infection with ePA. http://tinyurl.com/njqrbo3 ▲

Laura Tillman is 68 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

Published by the United States Adult Cystic Fibrosis Association, Inc.
CF Roundtable is printed on recycled paper.

IMPORTANT RESOURCES

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

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

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

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

ADA: To learn how the Americans with Disabilities Act (ADA) applies to you, contact the Disability Rights Education and Defense Fund (DREDF): Phone: 1-800-348-4232 http://www.dredf.org/.