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

How I Started With Weightlifting And My Motivation

By Garette Langmead

It's difficult to find the motivation to work out on a daily basis at 43 years old; add CF into the mix and it can be downright nasty. As difficult as it can be, I know if I don't work out at least five days a week it'll affect my health and daily life. I'll have increased cough, congestion, shortness of breath, body aches and pains, start feeling depressed, and overall feel lousy.

Getting started with weightlifting or any exercise program can be difficult. I can still remember my first weight set. It was my 13th birthday. My parents bought two five-pound dumbbells for me. It wasn't much weight, but for me, it was heavy. At the time, I was about 4 feet 8 inches and no more than 80 pounds. I could barely curl the weight eight times. My mom said she'd buy me a better weight set with a bench, once I could do 10 reps (repeats) of lateral raises (hands at your side, lifting weight up to shoulder height) with the five-pound dumbbells. Well, I could barely do three reps and at 13 years old I still lacked the motivation to get those 10 reps.

Fast forward two years—I wasn't much bigger, at most 5 feet and 90 pounds. All of my friends towered over me, all outweighed me by 30-plus pounds. I hated being small and I didn't know what to do about it. That's when I saw it. The magazine that would change my life, *Muscle and Fitness*. I was intrigued by the mere size of the guys in the magazine. I wanted to be them. That was the first time I was motivated to exercise and lift weights.

> "There are days I just don't have the drive or motivation to go to the gym, but my family keeps me motivated."

Luckily, I had a close friend who lived two blocks from me and had a weight set with a bench. It was the old school weight set; hollow barbell, cement weights wrapped in plastic and a narrow bench with the leg extension and leg curl attachment. He and I started lifting weights together the summer before our freshman year in high school. We used the magazine as a guide for our workouts.

Towards the end of that summer, we had a friend and his younger sister stop by while we were lifting weights in the carport. There were 70 pounds on the bar. Both of my friends could bench the weight 10 times. Then it was my turn, it was my first time benching

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INSIDE THIS ISSUE

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EDITOR'S NOTES

Autumn is here in all its glory. The leaves are turning lovely colors and are falling all over the place. This is the season to check with your doctor about whether you should receive a flu shot. The vaccines in recent years seem to do a pretty good job of protecting us. I hope this year will be another one where we can avoid catching the flu.

We have some changes in the USACFA Board of Directors. **Lisa Cissell** has had to resign, due to work constraints. Lisa had been a director for three years and was our secretary. She also worked on the outreach committee. We thank her for her time and hard work. She will be missed.

Jen Weber has been elected as the new secretary. Thanks for stepping up, Jen.

The newest director is **Ella Balasa**. You may read about her on page 17. Welcome Ella, we look forward to working with you.

As usual, this issue is packed with information that may help you cope with CF. It starts on the first page with **Garette Langmead** telling of his success with weightlifting and how it has helped him deal with CF.

In "Ask The Attorney," **Beth Sufian** discusses the changes in disability insurance being made by Medicare. **Aimee Lecointre** tells us we should learn to "sneak it in" when it comes to exercise. Be sure to check out "Active For Life" to see what she has to say.

Laura Mentch writes about her impressions of the 29th CFRI annual educational conference in "Notes From CFRI." There is more about that conference in "Searching For The Cure" by **Reid D'Amico**.

The focus topic of this issue is: Advocates And Advocacy. **Andrea Eisenman** tells of how her mother was her advocate and now she is an advocate for her mother. I tell of how my mother and husband have served as advocates for me, as I continue the focus discussion in "Speeding Past 50."

"In The Spotlight" focuses on **Victor Roggli, M.D.**, and his life with CF. **Dana Giacci Rogers** tells of how she planned for managing her CF before becoming a parent, in "Parenting." "Transplant Talk" finds **Paul Feld** updating us on how he is doing and where he is on his journey to kidney transplant.

The "Poetry Corner" finds another offering from **Linda Stratton** as she writes of her struggles to breathe. Be sure to check out the latest news on meds and research compiled by **Laura Tillman** in "Information From The Internet."

USACFA is accepting applications for the spring 2017 Lauren Melissa Kelly scholarships. Look at page 18 for information on how to apply. Be sure to check out that and the other announcements inside this issue.

Until next time, please stay healthy and be happy.

Kathy

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Diamond Sustaining Partners - In Memory of Lauren Melissa Kelly.

Information From The Internet...

Compiled by Laura Tillman

Opportunistic Pathogen Increasing In Cystic Fibrosis Patients

Burkholderia contaminans, a bacterium belonging to the Burkholderia cepacia complex (Bcc), is an opportunistic pathogen creating increasing concern for patients with cystic fibrosis (CF) in certain parts of the world. The predominance of B. contaminans infections has increased in Spain, Argentina and other Ibero-American countries. Frequently found in respiratory samples of CF patients in those areas, the bacteria often leads to debilitating lung infections and high risk for developing deadly septicemia — but little more was known. The team of researchers performed a genome-wide comparative analysis of two isolates of the bacteria from the sputum and blood of a female CF patient in Argentina. The research team found that samples of the bacteria isolated from the patient were genetically very different from one another; more specifically, the two isolates differed by more than 1,400 mutations. Data showed that there were significant changes in the expression of bacterial genes involved in the virulence (severity), motility (movement) and chemotaxis (the process by which bacteria move by sensing their chemical environment). The scientists also detected changes in the expression of genes that are activated in stressful conditions for the bacteria such as low-oxygen, and that encode for stress-related proteins. Finally, the researchers observed some changes in the expression of genes responsible for the biosynthesis (process of new substances developing within the body) of anti-fungal and hemolytic (red-blood-cell destroying) compounds. Researchers found that there were two distinct types of B. contaminans that coexisted in the body of the patient and entered her bloodstream. They found that B. contaminans isolates can hold a high genomic diversity, potentially resulting in different bacterial virulence.

http://tinyurl.com/zzm9bgd

Chemical Exacerbates Bacterial Infection, Important For Cystic Fibrosis

Interaction between bacteria naturally found in the lungs and disease-causing bacteria may make it harder to clear the pathogen from the body. Preclinical findings explore the relationship between 2,3-butanediol, a chemi-

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LOOKING AHEAD

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

Autumn (current) 2016: Advocates And Advocacy.

Winter (February) 2017: Using Non-traditional Medicine Or Treatments. (Submissions due December 15, 2016.) Have you tried or are you interested in trying any non-traditional medicines or treatments? Do you have any good advice or past results that you can share with the readers?

Spring (May) 2017: Traveling With CF. (Submissions due March 1, 2017.) What experiences have you had traveling with CF? Are there any pitfalls that we can avoid? Do you have any tips that you can share? Please tell us of your experiences.

Summer (August) 2017: Problem Solving With CF. (Submissions due June 15, 2017.)

ASK THE ATTORNEY

Breaking News: An Analysis Of The New Social Security Rules For Cystic Fibrosis

By Beth Sufian, J.D.

In this column we explore views on the new Social Security medical eligibility criteria for people with CF. The new rules went into effect on October 7, 2016. As Director of the CF Social Security Project®, there are certain insights I want to share with my community that relate to the new rules.

Most important, the CF Social Security Project will continue to represent individuals with CF in an initial application or in a Social Security Continuing Disability Review. The CF Legal Information Hotline® will continue to provide information about Social Security benefits, insurance, employment and education to the CF community. The Hotline is sponsored by a grant from the CF Foundation and can be reached at 1-800-622-0385 or by e-mail at CFLegal@sufianpassamano.com.

Nothing in this column is meant to be legal advice about your specific situation and is meant only as information. Meeting one of the medical criteria listed below does not guarantee eligibility or continued eligibility for SSI or Social Security Disability benefits.

The Social Security Administration (SSA) has changed the medical eligibility criteria it will use to determine if an applicant with CF is eligible for benefits. The Social Security medical eligibility rules that have been in effect for more than 20 years have allowed an applicant with CF to medically qualify for Social Security benefits if the applicant had either: (A) low FEV_1; (B) six physician interventions in 12 months; or (C) use of an inhaled or home IV antibiotics to treat a persistent pulmonary exacerbation once every six months in the year prior to applying for benefits. **Starting October 7, 2016, these rules will change.**

One of the most significant changes is the old §3.04B will be deleted from the rules. This section previously allowed a person with CF who had six physician interventions in the 12 months prior to applying for benefits to meet the medical criteria for benefits. Section 3.04B was a flexible standard. Many applicants with CF became eligible for Social Security benefits under this section. Unfortunately, this section will be entirely deleted on October 7, 2016. According to the new rules, a**ny cases pending on October 7, 2016, will be determined using the new rules.**

Another significant change is the removal of the old §3.04C from the rules. The old §3.04C provided that a person using inhaled or home IV antibiotics (for any amount of time) to treat a persistent pulmonary infection once every six months met the eligibility criteria. In practice, Social Security also required a showing of significant daily treatment time. Many people with CF qualified for benefits under the old §3.04C. Unfortunately, this criterion also will be entirely deleted.

BETH SUFIAN

It is important to understand that even if a person does not meet one of the new criteria the law still allows Social Security to approve a person who shows his medical condition is medically equivalent to one of the listed medical criteria. However, a determination based on medical equivalence is usually made by an Administrative Law Judge. Currently it takes an average of 522 days to have a hearing before an Administrative Law Judge in the United States.

The new rules retain low FEV_1 as an eligibility requirement, but this requirement has been substantially reformulated and is now based on age and gender. In addition, there are certain FEV_1 values for those who are under the age of 18. There is also a column of FEV_1 point values that are higher than all the other columns. It is unclear if those point values are to be used for a person who files an application for benefits when they are 18 or 19 years of age and do not have a decision from SSA before they reach the age of 20. A request for clarification on this issue has been made to SSA. Once the CF Social Security Project receives an answer from SSA, there will be a blog post on the *CF Roundtable* blog regarding the answer.

Social Security looks at medical records within the 12 months prior to

the filing of an application. Under the new rules a person may be eligible in six other ways:

1. the person has had at least three hospital stays in the year prior to applying for SSA benefits with at least 30 days between each set of two stays; OR

2. the person is treated for a spontaneous collapsed lung requiring chest tube placement; OR

3. the person has respiratory failure requiring use of invasive ventilation or noninvasive ventilation with BiPAP or a combination of both treatments, for a continuous period of at least 48 hours or a continuous period of at least 72 hours if following surgery; OR

4. the person has an embolization for hemoptysis; OR

5. the person has low oxygen twice in a 12-month period at least 30 days apart. The values given depend on the altitude where the person is living. For example, if the person is living at less than 300 feet above sea level, the oxygen value will need to be 89% to meet this criterion. If the person is receiving supplemental oxygen and therefore has higher oxygen level it is unclear if the person could meet this criterion.

The sixth way to qualify requires a combination of any **two** of the following:

- 1 Pulmonary exacerbation requiring 10 consecutive days of IV antibiotic treatment (the IV use must be 30 days apart to count as two separate episodes);
- 1 Episode of hemoptysis (more than blood-streaked sputum) requiring hospitalization of any length;
- 1 Weight loss requiring daily supplemental enteral nutrition via gastrostomy (feeding tube) for at least 90 continuous days or parenteral nutrition via a central venous catheter for at least 90 consecutive days;
- 1 CFRD requiring daily insulin therapy for at least 90 consecutive days. It is unclear if the person will have to show that the CFRD is uncontrolled or if controlled CFRD will allow a person to meet part of this section of the medical listing.

> *"It is helpful if medical records contain information about daily limitations that prevent a person from working full time."*

The new rule has a welcome addition addressing people who are up to three years post-lung transplant. It adds a new section establishing a presumption of disability of up to three years post-lung transplant. The section is found at Section 3.11 of the Social Security Listing and reads "Consider under a disability for three years from the date of the transplant; after that evaluate the residual impairment." It is unclear if merely showing that a person is less than three years post-lung transplant will be enough to be eligible for benefits or if the person will be required to provide medical records showing that the person is physically unable to work full time.

This new section does **NOT** mean the person will lose benefits at their three-year post-transplant anniversary.

After a person is three years post-transplant, the person may receive an SSA Continuing Disability Review. The person will then have to show SSA medical evidence that supports a finding the person is unable to work full time due to his or her health issues.

The importance of medical records when applying for Social Security benefits or completing Continuing Disability Review should not be underestimated. Visiting a CF Center for quarterly visits is important. SSA questions the credibility of a person who asserts he is unable to work because of CF health issues but sees his or her physician only twice a year. The new rule changes necessitate having medical records that clearly discuss daily issues a person has that prevent the person from working full time.

While there is no exhaustive list of what should be included in a person's medical record, it is helpful if medical records contain information about daily limitations that prevent a person from working full time.

Information such as the time it takes to perform each breathing treatment done in the morning and each treatment done at night, the number and duration of airway clearance treatments, the need for naps during the day, issues with coughing up blood or digestive issues, issues with memory, concentration or stamina. Any information that supports a finding that the person cannot work full time is helpful.

There is no guarantee that including such information in a person's medical records will result in an approval for benefits. It can help to have detailed information about a person's medical condition in his or her medical records.

The CF Social Security Project handles cases for people with CF from around the country and so is able to assess trends in how local Social Security offices are applying the new medical criteria. Watch the *CF Roundtable* blog in the coming months for updates on how SSA is interpreting the new rules. Also feel free to contact the CF Legal Information Hotline® with any questions regarding Social Security benefits. ▲

Beth is 51 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.

SPEEDING PAST 50

An Advocate Can Make Life Easier

By Kathy Russell

Autumn is here at last. The weather is turning cooler and the leaves are beginning to fall. (Maybe that is why so many people call this season *fall* instead of *autumn*.) This is the time that I like to can tomatoes and make pickles and relish for my pantry. I am so happy that I still am able to preserve foods in these ways. It takes a lot of my energy, but I am happy to use it that way.

At any rate, autumn brings a new set of concerns for us. Many of us get a "flu shot" to help us prevent a bout of influenza, aka "the flu." I don't know how flu and other respiratory bugs affect you, but I know that they cause me great concern. It seems that I have a much harder time getting over infection from such bugs as the years go by. I avoid catching everything that I can. (That is because it is much easier to avoid them than trying to recuperate from having caught one of these bugs.) My pulmonologist recommends that I get the extra strength flu vaccine. Of course, I am old and that is what is right for me. Younger people may need a different vaccine. Ask your physician what he or she recommends.

Autumn is also a good time to be more vigilant about using hand cleaner when dealing with anything that others may have touched. My supermarket has disinfectant wipes near the shopping carts so that I can wipe the handle of a cart before I use it. Believe me, I use them. I also am more likely to wear gloves (just regular gloves, not vinyl or hospital type) when I go away from home during autumn and winter. I really try to avoid other people's bugs. The first thing I do when I return home is wash my hands. I don't want to bring anything home, if I can avoid it.

Looking out for ourselves is so important, but it can be difficult. Sometimes we are not able to look out for ourselves. We just don't have the energy that it takes to look out for ourselves. Then what should we do? I think we should get someone else to assist us.

When I saw the Focus topic for this issue: Advocates And Advocacy, I decided to look up the definitions. I found that an advocate "pleads on someone else's behalf." Advocacy is "public support for or recommendation of a particular cause or policy." How does this relate to us? We have so many interactions with others that use up more energy and take more time than we may be able to spare. Pleading our case with a medical facility or insurance company can be exhausting and it can take hours or even days to get satisfaction. Sometimes we just give up. That is not a good resolution of a problem.

One way to handle such problems is to have someone act as an advocate. We all know that social workers can and do act as advocates, in certain situations. However, most of us don't have a social worker on hand all the time. We have to find someone else who will advocate for us. That person may be a family member or a special friend.

> *"Paul handles all the business that I don't want to bother with or don't have the energy to do."*

KATHY RUSSELL

When I was young, my mother was my advocate. She had to fight to get me properly diagnosed. My docs had trained before CF was identified so they weren't really cognizant of what it was. Mother fought for everything that I needed and made sure that the docs treated me when it was necessary. She scheduled my appointments and saw to it that I got to them. She carried out any treatments that were prescribed. She stayed up with me when I was having trouble breathing and when I had high fevers. She always was there for me. Thanks, Mother.

When I was about 16, I started doing more of my scheduling and care for myself. I felt it was time for me to get used to taking care of myself. I wanted to be ready to fend for myself when I went away to school and when I lived on my own. I did okay with that until I was much older.

In more recent years, my advocate has been my husband, Paul. I have requested that Paul be able to handle all of my business with Medicare and with all of my doctors, insurance companies and medical facilities. I have signed authorizations to allow him to have access to all of my medical information and to speak on my behalf. This has saved me countless hours of irritation.

Paul handles all the business that I don't want to bother with or don't have the energy to do. I really don't like to talk on the phone. I really, really don't like to *wait* on the phone. I am not a patient person when it comes to "holding" on a phone. It isn't just because of the music that most phone lines blare while I'm holding. (Of course, that does contribute to my displeasure.) I just resent having to spend time holding. Paul is much more patient than I am. He holds and listens and handles whatever business there is to be done. I know how fortunate I am to have someone to handle all of these things for me.

Another place where Paul's advocacy makes my life easier is regarding prescriptions. On the first of every month, Paul and I review our prescription medicines to see if we need to order anything. We get our prescriptions via mail order from the pharmacy that our insurance dictates. Paul makes sure that we have enough on hand that we don't run out. If there are any questions or problems, like when they don't want to provide what the doctor has ordered, he handles them. That is a big load off my mind.

Paul serves as an advocate with my medical appointments. He goes with me to my doctor visits and listens, as well as takes notes. He often catches things that I may have missed. He also takes along a list of questions or comments that have arisen since my previous appointment. Our family doctor and my pulmonologist are accustomed to this way of doing business and seem to have no problem with it. It takes a lot of pressure off of me to have to remember everything.

While I am seeing my doctor, if I am asked how I am doing and I reply that I am doing okay, Paul may step in and say that I am not or that I have been complaining of... or unable to do... or some other comment. It is so easy for us to get in the habit of just saying, "Okay," when we should say that we have been feeling crummy or having this or that problem. I think this comes from a combination of trying to be polite, not wanting to have to explain how we actually feel and the need to be the "good little patient." Our advocates can call us on this behavior and remind us of how we have been feeling – *really*. Our docs appreciate getting the "real story."

When I go to either admitting at a hospital or the emergency department, Paul makes sure that all of my information is correct and that everyone listens to what I say. He also amends or corrects my comments when I say something incorrectly. When I am not feeling well, I am prone to make errors. It is so easy for people in those hospital departments to get so accustomed to people's stories that they don't really listen. Advocates can make sure that they do pay attention and that they get it correct.

When I am unable to make myself heard or understood because of how ill I am, Paul is able to make sure that my caregivers know how I am and what I need. He looks out for me and helps me to get the best possible care that I can. I appreciate all that he does on my behalf. Thanks, Paul.

I think that having an advocate can make one's life easier. As a rule, when someone is ill they don't have the normal stamina to do things. Having to argue with an insurance company or some other healthcare provider takes a lot of energy. That energy is needed for getting well. An advocate can help to get you up and back at your normal life in less time than if you have to do it all by yourself. I strongly recommend that you find someone to be your advocate and then – let them help you. It can make your life easier.

Stay healthy and happy. ▲

Kathy is 72 and has CF. She is Managing Editor of CF Roundtable. *Her contact information is on page 2.*

Cystic Fibrosis Mothers

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

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

If you would like to join our highly active and friendly community, please message Emma or Alison on one of these links:

https://www.facebook.com/emma.harris.16
https://www.facebook.com/alison.w.smith.7

PARENTING

Balancing CF And Parenting

By Dana Giacci Rogers

The longer I have been a parent the more I realize that a reasonable routine can create success for all. For most first-time parents the primary advice is to find time for yourself. However, not all parents have CF and CF comes with its own set of time and energy requirements. When you have CF, carving time out for yourself as a parent means not skipping morning treatment because the kids are late for school, and making every treatment every day a requirement. Allowing hospital stays or cleanouts to happen when needed. Or for the love of everything holy, just getting enough sleep because your body is already taxed enough! Carving out time for yourself as a CF parent isn't just good advice, it is a skill for survival. It is with this thought in mind that I would like to discuss some tips or suggestions for a successful transition into parenthood when you have CF. I can speak only from my experience but, with three kids, balancing starts to become second nature.

Before my first pregnancy, my husband and I looked at every possible angle of CF life and tried to focus our energy on what that would mean for a newborn, for a toddler and just for young kids in general. We tried to devise a plan for handling each situation that arose. Life being what it is, you can't plan for everything and even the best laid plans go out the window at times. But having a plan in place can relieve stress and allow what needs to happen to happen. Since my husband works, we needed to decide on child care. At that time, I was still in school and we had the discussion of whether both a career and children were reasonable with my CF. Perhaps this is the question to start with. Demands of children and CF and a career are all different and all important. It's good to know your limits and what you can handle so you don't make yourself sick in the meantime.

From there we asked ourselves, what about hospital stays and cleanouts? At that point in my life I was dead set against having a port placed. I wasn't ready and PICC lines were so easy. Three weeks and done, no maintenance, no blood clots, no worries. However, my veins started to refuse picc placements long before pregnancy. It got so difficult that every placement had to happen in interventional radiology with a one- to two- night minimum stay, even if I decided to do home care IV afterwards.

So the plan we made was to have a port placed and do cleanouts at home. That way planning around the hospital with a small child happened less, although it wasn't entirely eliminated. But less was good. That way I could stay home with my kids even when I needed extra IV meds and there was no separation anxiety to worry about for them or me. This is just how it worked for us. I hear some CF parents like to just schedule hospital stays so they get a restful break. As long as you have the child care in order, that is certainly an option too. In the end if you have thought about a plan, then you have made time for yourself when you will need it, and that is a good start.

> *"Carving out time for yourself as a CF parent isn't just good advice, it is a skill for survival."*

DANA GIACCI ROGERS

Now, what about daily treatments? This one is harder to plan for. Each stage of a child's life brings a completely different routine and needs along with it. As long as you are flexible and you are diligent about finding the time with each new stage and each new schedule, it can be accomplished. I learned this lesson best with my stepkids, before the arrival of my daughter. I used to take my stepson to school first thing in the morning, but my stepdaughter was in afternoon kindergarten. So I would drop him off and we would head back to the house. I allowed my stepdaughter to watch morning cartoons and this allowed me to have quiet time in order to do my morning treatment. As long as I had my morning treatment started by 9:30, there was still time for the park and lunch before school started. The evening treatment always was after saying good night.

That worked out for a while.

Then, after my daughter was born it looked a little different still. I would drop my stepkids off at school and the baby and I would head back to the house, just in time for morning nap at about 10 a.m. or so. I would have time for morning treatment. Then with the arrival of the twos, morning nap was eliminated altogether. Now what? I wondered. Admittedly, I do a very late morning treatment on a regular basis that correlates with afternoon nap. But soon she will be old enough to stay out of trouble like my stepkids before her and perhaps I can transition back to morning treatments like I did with my stepdaughter. Either way I have missed very few treatments as a CF parent, which was the one thing my doctors and family seemed most concerned about. I am diligent about finding my moments and instead of getting a traditional parent break, my CF gets taken care of.

Perhaps try to imagine what you do on a daily basis... how many treatments or how long they take. Can you fit them into nap times? Or do you need a partner tag team treatment time? Or perhaps it's just easiest to wake up early and do treatments before anyone wakes up and go to bed late doing treatments after everyone is in bed. I did that for a number of years with my stepkids. That is harder but there is absolutely no more worry about when treatment will fit into a busy morning or evening schedule. These are good scenarios to think about because, no matter what, you will always have daily maintenance to do and you can do it!

Finally, we asked ourselves, what about lung transplant? For something so big and unknown until we go through it, I knew I couldn't make educated guesses as to what that might look like. The only thing I knew was that the older our kids are before that day comes the better it will be for everybody. That was my personal opinion and my husband's too. So we started planning our family immediately at that moment, so that when the time came our kids would hopefully be a little bigger, a little more independent and a little more CF-wise.

In the end, all things are possible. Being a parent is something I would never change for the world. I am extremely fortunate that I took that leap. I am grateful for the person I took it with. I appreciate all my time with my kids. Although as they are older and have three to four activities at once, things can get a little hectic, but I still enjoy watching baseball in the spring and soccer in the fall. Every moment of your kids' lives or soon-to-be lives is a great gift. Even greater that you have so much wisdom about living life to the fullest to teach them. Just remember that being a parent with CF – longevity is the goal. Both you and they deserve it. So find the time for yourself in a very new and possibly difficult kid schedule. Strive to optimize moments where treatments could be done. Look at running around with your kids as airway clearance and do it for a little longer. In the long run it is best for all of you.

Please feel free to e-mail me if there is a particular topic about CF and parenting you would like to read about. If I do not have personal experience with it, I will do my best to search out other CF parents who might. Also feel free to e-mail me if you would like to share your CF and parenting story.

Stay healthy and happy. ▲

Dana is 27 and has CF. She lives in Troutdale, OR. You may contact her at: dgiacci@usacfa.org.

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cal produced by the bacteria that naturally live in the lungs, and P. aeruginosa. The interaction between the two components yields an infection far more harmful than P. aeruginosa would create alone. The findings also demonstrate that a concoction of naturally occurring bacteria, known as a microbiome, exists in the respiratory tract — and that it can play an important role in influencing health. According to the authors, it is important to better understand how P. aeruginosa infection settles in the lungs and how the bacteria interacts with other bacteria to find ways to effectively treat the infection.
http://tinyurl.com/zj6rnk2

AND

http://tinyurl.com/h64ksog

Cystic Fibrosis Inflammation May Be Caused By 'Helpful' Bacteria

The abundant mucus in the lungs of cystic fibrosis (CF) patients helps certain types of bacteria, called anaerobes, to survive and, in turn, support the survival and proliferation of more dangerous opportunistic pathogens like Pseudomonas aeruginosa, which otherwise would not find the nutrients they need in mucus-lined airways, a study reported. This symbiotic relationship appears to underlie the inflammation that characterizes the disease. The study suggests that, although certain bacterial species cannot use lung mucus as a viable substrate, other types find mucins, the major constituent of mucus, a large and rich pool of essential nutrients like amino acids (nitrogen) and sugars (carbon). The study reveals that a subset of CF microbiota is capable of fermenting mucins for carbon and energy which, in turn, can support

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TRANSPLANT TALK

So Far, It's Been A Good Run, But...

By Paul Feld

It's been a long time, but my fingers and mind are getting antsy and it's time to write again. Truth be told, this is a mental health issue and this is my best way to deal with it. If you have been a long-time reader of *CF Roundtable,* you may have read some of my life experiences before. If not, I'll provide a brief update. I was diagnosed with CF at 19 years of age, in 1976. I had a bilateral lung transplant in 2004, participated in three U.S. Transplant Games and won a couple medals. Next year, I hope to reach the age of 60 in May. I have been close to or near death a couple of times, probably once in each of the last four decades. Each time was a horrible experience, but somehow I fought through it, mostly because I felt I could. I am now in that situation again. This time, however, the fight is long and not immediate and I'm not sure I have the will to conquer as I have in the past. Thus, I have to write.

I am in my seventh year of stage-4 kidney failure, with my kidneys now functioning at 13-15%. I have been very blessed to have survived this long in stage-4, and my loving family has certainly played a big part in getting me here. I have been on a kidney transplant waiting list for about three years, and was recently told by my healthcare team I am now fourth on the list. A couple weeks ago I had a catheter placed in my abdomen to begin the process of peritoneal dialysis. It will be another month or so before I can actually begin dialysis as my catheter hole heals.

So many things go through my mind daily in my current status. First and foremost, I think about death. My father passed just a few months ago, along with my aunt, his sister. I was with them both almost every day for the last month of their lives. While they both went peacefully, with the exception of a couple days, the memories are still very vivid and, to some extent, haunt me. Mostly because the last week of their lives, they were morphine-sedated and their bodies slowly wilted away. It's just not the way I want to leave my family.

I went on disability for the first time in January of this year. My whole life I have battled CF, I was able to work full time and carry my own weight in society. Things have changed very quickly and I now depend on SSDI and a small pension to make ends meet. Fortunately, I still have my primary health coverage through my wife's employer, then Medicare. More importantly, I now depend on my family to help me take care of my own health. That has happened only once before, for about three months post-lung transplant. My wife, Kristi, and my sister, Julie, are getting dialysis education right along with me. I feel as though I am so impeding on their lives and want to be independent again. I am fairly certain most aging adults get the same complex, but most of them are in their 70s or 80s, not 59.

A few months ago, for some reason, my health went downhill very quickly. In March I could keep up my routine of walking at least one and a half miles every day. Within a couple of weeks, I could not even walk to my mailbox and back (about 30 yards) without stopping to rest. My lungs are fine, but my legs just don't want to move, no matter how hard my mind wills them to. They just turn into spaghetti. After five minutes or so, I can go another 30 yards. It's as if my muscles atrophied overnight. This is what causes me most concern, as I wonder what will fail me next.

Finally, I have unending dates with my MOHS surgeon, who has, over the last five years, carved out about two

PAUL FELD AND HIS SISTER, JULIE LAUGHLIN.

dozen trails on my scalp and face. Ninety-five percent of the skin cancers have been squamous cell, but it takes only one to take a deeper root and enter my brain. If you want to meet the real "Young Frankenstein," lets meet someday. My surgeon has done a wonderful job, considering her challenges, but a closer look provides a good roadmap of the interstate highway system in our country.

> *"I have been very blessed to have survived this long in stage-4 [kidney disease], and my loving family has certainly played a big part in getting me here."*

Through my life, I have generally been a pretty positive person, mostly taking care of myself independently from the rest of my family and world. Meeting me on the street, you'd walk right by not knowing anything was amiss. Now, everyone notices something and the attention is focused on me, not something my personality appreciates.

This article doesn't have a positive or negative ending, but it did allow me to put my thoughts on paper, and that in itself was healing. To my many CF friends who read *CF Roundtable*, I miss you daily and often wish we lived closer. I will miss the CFRI retreat this year, where I usually spill my heart to 40 or so others who may be sharing their same concerns and thoughts. Know that I'll be invisibly following you around this year, laughing and crying as you go. Enjoy your week, and continue to lean on each other. ▲

Paul is 59 and has CF. He was a director of USACFA 14 for years and served as President. He lives in St. Peters, MO, with his wife, Kristi. You may contact him at: feld.paul@gmail.com .

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the carbon demands of other respiratory pathogens in co-culture. Fermentation is the process through which bacteria produce molecules for their maintenance in environments of low oxygen, such as what occurs in the lower respiratory tract (trachea, bronchi and lungs). These low-oxygen bacteria are called anaerobic, or anaerobes. Researchers also observed that products indicative of mucin fermentation were abundant in CF patient sputum and that P. aeruginosa expressed several genes needed to use these products. Together, the observations support the potential role of fermentative metabolic reactions in the development of CF lung disease. These results suggest that the nutritional dynamics among bacteria contributes to the development of CF lung disease. This finding may also help in the design of new therapeutic strategies for the management of disease progression.
http://tinyurl.com/h8vgfe6

Vertex Ends Trial Of Cystic Fibrosis Drug Combination

Vertex reports that one of its Phase 3 clinical trials looking at the combination of VX-661 with ivacaftor to treat various subgroups of cystic fibrosis (CF) patients has been stopped. The study being terminated involved patients with one copy of the F508del mutation and one copy of a mutation that results in minimal CFTR protein function (F508del het/min). Three other Phase 3 studies involving VX-661 plus ivacaftor will continue. One study is assessing the drug combination in patients with two copies of the F508del mutation, a second in patients with one F508del mutation and one residual function mutation, and a third in patients with one F508del mutation and one mutation that results in a gating defect in the CFTR protein. VX-661 is a CFTR corrector that modulates the folding and trafficking of CFTR protein. Ivacaftor (Kalydeco) is a CFTR potentiator that improves the transport of chloride. It is approved for the treatment of CF patients with CFTR mutations: G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, S549R or R117H.
http://tinyurl.com/hv74vnq

NovaBiotics Announces New Data On Cysteamine (Nylexa™) As An Antimicrobial Resistance Breaker In Multi-Drug Resistant Bacteria

NovaBiotics, Ltd., presented data outlining how Nylexa™, its parenteral formulation of cysteamine in early stage development, improves the antimicrobial efficacy of antibiotics and reverses multi-drug resistant (MDR) infections. NovaBiotics first discovered cysteamine as a multi-active antimicrobial in its research into cystic fibrosis (CF) therapy and the clinical trials of its novel CF therapy Lynovex®. Results show that cysteamine reverses resistance to clinically important antibiotic classes such as aminoglycosides, fluoroquino-

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LANGMEAD continued from page 1

that weight. I laid on the bench, picked up the weight, brought it to my chest and no matter how hard I tried I couldn't push it off my chest. Next, my friend's little sister decided to attempt the weight. She benched it three times. It was an extremely embarrassing moment for me. Here's a girl who was two years younger than I and she could out-lift me. My friends razzed me for months about how she was stronger than I was. I couldn't use having CF or being smaller than her as an excuse. At that moment, I was motivated more than ever to get bigger and stronger. I didn't want that happening again—ever.

During high school, I took weight training every year. Each year I noticed that I was growing and getting significantly stronger. I was really strong for my size. By the time I graduated high school, I was 5 feet 6 inches and 120 pounds. I was still a little guy but I could out-lift most of my friends who outweighed me by 100 pounds.

I continued to lift weights through college, but at that time I really didn't have a lot of motivation to exercise and lift weights because I was working and going to school.

GARETTE LANGMEAD COMPETING AT THE 2007 WABDL WORLD COMPETITION.

I was just lifting to maintain my health and keep my lung function up so I wouldn't end up in the hospital. I still gained strength and size through my college years even though my workouts were limited due to time and the fact that I was eating on a budget. By the time I graduated college my weight was up to 135 pounds.

In my late 20s, I started working out at a new gym. The first week I was in the gym, a guy came up to me and asked if I had ever competed in powerlifting. I said, "No." He asked how much I weighed and said I was strong for my

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lones, macrolides, folate pathway inhibitors and beta-lactams. NovaBiotics also presented data to show that cysteamine reverses MCR-1 and other forms of colistin resistance. Lynovex®, in addition to standard of care antibiotic therapy, reversed single and multi-antibiotic resistance in P. aeruginosa and S. aureus isolates.

http://tinyurl.com/zutn6lk

Proteostasis Therapeutics Issues Mid-Year Corporate Update On Cystic Fibrosis Pipeline Programs

Proteostasis Therapeutics' lead drug candidate is PTI-428. It is the first genotype-agnostic, disease-modifying agent to be tested in the clinic for cystic fibrosis (CF). PTI-428 is a unique modulator (amplifier) of the cystic fibrosis transmembrane conductance regulator (CFTR) protein that has shown a consistent positive effect on CFTR mRNA and protein activity in vitro and in preclinical studies when used in combination with existing treatments. Preliminary data in healthy volunteers suggest a dose-linear pharmacokinetic drug profile that supports once-a-day dosing. Data confirmed the expected and dose-dependent relationship between PTI-428 exposure and the magnitude of CFTR mRNA increase. A single dose of PTI-428 can increase CFTR mRNA expression by approximately two-fold in those healthy volunteers. No safety concerns have been identified to date based on reviews of vital signs, clinical exams and hematology lab values. In vitro and in vivo animal studies with PTI-428 have shown that a two-fold increase in CFTR mRNA may lead to a potential doubling of lung function improvement on top of the existing standard-of-care therapies. Additionally, Proteostasis Therapeutics have further advanced preclinical development of PTI-801 (corrector) and PTI-808 (potentiator) molecules which, when combined with PTI-428, are the active components of

size. Then he asked if I'd be interested in competing with his powerlifting team. I said, "Sure." I needed something to keep me motivated in the gym. Most days I just felt like I was going through the motions to stay healthy, but I didn't have a passion for lifting anymore. I figured being a part of a team would keep me motivated.

Later, I found out that this guy owned the gym and was a world class powerlifter with multiple world records. For the next year, I trained with the team. Then I competed in my first powerlifting meet in March 2002. I set two state records in bench press and three state records in deadlift for the 148-pound weight class. I was hooked! I found my new motivation to keep going in the gym. Over the next 10 years, I set 30-plus records and competed in two world competitions where I took third place in 2002 and first place in 2007.

I haven't competed in the last four years, due to some changes in my health. However, I still train five to six days a week. I train with my teenage boys and enjoy teaching and watching them grow. There are days I just don't have the drive or motivation to go to the gym, but my family keeps me motivated. They push me when I'm not able to push myself.

I've had some significant hospitalizations over the years. There were two hospitalizations about 10 years apart that brought my FEV_1 from high 70s into the low 20s. Both hospitalizations were 30-day admissions and I went home with my FEV_1 in the 20s.

After every hospitalization, I'd come home and just want to rest. Heck, I would get short of breath just walking to the bathroom. I wasn't even considering walking into a gym. Luckily, I've always had someone to push me to get me back into the gym, even if I didn't want to go. My son, Noah, has pushed me to get back in the gym every time I come home from a hospitalization, since he started training with me three years ago. That push has brought my lung function up every time and keeps me motivated. I've been blessed to have great friends and family around me to keep me motivated to exercise and fight CF.

Through the years I've had many motivators to keep me weightlifting: a fascination with a magazine, a girl outlifting me, competing in powerlifting and now, staying healthy for my family. My passion for weightlifting started with that first *Muscle and Fitness* magazine I read. Finding the type of exercise you're passionate about and what motivates you to exercise can be difficult, but it's necessary to fight this terrible disease. ▲

Garette is 43 and has CF. He was diagnosed at five months of age. He's married and has six kids; biological triplets and three stepchildren. He worked 18 years as a Registered Respiratory Therapist and a Registered Polysomnography Technologist and now works as a Certified Personal Trainer. He's been a competitive powerlifter since 2002, had over 30 powerlifting records in bench press and deadlift, and is a team trainer/co-captain for the Northwest Barbell Club. He enjoys spending time with his wife and six kids, camping, hiking and working out with his four boys.

He offers free personal training to anyone with CF. If you are interested in a training program or have any questions, you can contact Garette at: innerstrengthtraining@yahoo.com

the proprietary triple combination therapy PTI-NC-733. In the Ussing chamber assay, PTI-NC-733 achieves approximately 100 percent of normal CFTR levels, which could translate into a more clinically meaningful benefit to a broader set of mutation classes than current or pipeline therapies.
http://tinyurl.com/h7bnw5c

CF Drug Candidate AB569 To Begin Production For Human Trials

Arch Biopartners and Catalent Inhalation, a division of Catalent Pharma Solutions, have entered agreements to start manufacturing AB569 for human trials in patients with chronic and antibiotic-resistant bacterial lung infections. Patients with cystic fibrosis (CF) stand to benefit most. AB569 was invented by Daniel Hassett, PhD, professor at the University of Cincinnati College of Medicine. Last year, AB569 received orphan drug designation in the E.U. and U.S. for the treatment of CF. The drug particularly targets the bacterium Pseudomonas aeruginosa of which the mucoid form is difficult to treat with traditional drugs. Arch scientists believe that the two active ingredients in AB569, acidified nitrite and EDTA, boost the actions of each other so that the final result is better than expected from the two substances separately. The drug will be developed as an inhaled solution.
http://tinyurl.com/hm6o5yw

Phage Therapy Against Antibiotic Resistance In CF Gets New Patent

Bacteriophage-based therapy is a potential therapeutic tool for the treatment of bacterial infections. Bacteriophages are viruses able to infect and replicate within bacteria. The new patent covers the treatment of Pseudomonas infections through the sequential use of a bacteriophage (phage) therapy, followed by an antibiotic to which the bacteria were previously resistant. During the first stage of

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FOCUS TOPIC

ADVOCATES AND ADVOCACY

Advocacy And Its Importance In CF And Beyond

By Andrea Eisenman

Growing up with cystic fibrosis (CF), for me, meant constantly going to doctor visits. Or rather, CF clinic visits, where I saw the nurse coordinator, pulmonologist, dietician and a social worker, after an x-ray and a PFT. During those clinic visits, I would be asked by everyone mentioned above, How was I doing? To which I would answer, "Okay." This made my mother crazy when she came with me, because I was not "Okay." I had chronic lung infections for which I took prophylactic oral antibiotics and when things got bad, such as increased shortness of breath and fatigue, I was put in the hospital (this was before home IV therapy was an option) for IV treatment on double antibiotics.

Even when I was admitted to the hospital in my late teens, because I was short of breath and completely congested, the attending physician would do my intake and ask, "How are you today?" Again, I would say, "Good, how are you?" Like we were two friends meeting on the street. That is when my mom would take over and tell the doctor what was really happening, just as she did at clinic.

I am not saying this was good on my part. I think I was in severe denial and just didn't want to deal. I was angry at being different from my peers and rebelled. But, eventually, I learned to deal with it because my mom was not always with me at doctor appointments or when I had to be admitted through the ER for treatment. It took me a while, in my 20s and 30s, when I was finally living independently, to understand that communicating with one's doctors was of utmost importance. And that working with them on my own care was like being part of a team. I know, I am a slow learner.

Now it is second nature to me to tell the doctors what is going on and make sure they know what I feel is the issue and what might help me. And now, with my lung transplant, I have many different specialists I see on an almost monthly basis. Of course, I let them test and examine me, but we are both working toward the same goal: getting me healthier and not bothering them so much!

ANDREA EISENMAN

Part of my problem was, when I would get sick, I had a gut feeling that I had to be admitted, especially in my 20s, but wanted to avoid it at all costs. As we know with CF, it only gets worse without treatment. Then, when I started to get sick more frequently and needed antibiotic IVs on a regular basis, I stopped fighting it because I knew waiting was detrimental to my health. I had to be more compliant and therefore would not wait until I had to be admitted but would ask for IVs earlier and do them at home with my port.

Through many years of having this chronic disease and then almost dying from it in end-stage CF, I finally learned many things about talking to doctors and advocating for myself.

I felt my doctors appreciated my new approach to being more compliant and advocating for what I wanted. Initially my doctor would not allow me to get a port placed because he felt they were unsafe. But then when he saw how well I did and how I stopped fighting him on IVs, he had almost all of his patients get them. It also allowed me to do my IVs at home and not in the hospital, which was usually a bad experience.

Here are a list of some things that may be helpful to others:

1. Don't be afraid of your doctor(s). I see people become intimidated easily and not tell them what is going on. Whatever you feel is going on is valid and will help your doctor or nurse with your diagnosis and hopefully a fix.

2. Don't be afraid to remind your doctor of something critical in your care. If you feel something was overlooked, just say something. They see many patients and it happens.

3. Make a list to discuss at your doctor appointment. I still do this about two weeks before my transplant

checkup. Many things come up and I find I can be so distracted and tired from waiting a while to see my doctors, I forget what I had wanted to discuss if it is not an acute issue.

4. Bring someone with you to appointments. At times we get bad news or suspect we will, or things just become overwhelming. It can be good to have an extra pair of ears. It is also good if they can take notes for you so you can concentrate on talking to the doctor.

5. Get your doctor's or nurse's e-mail, if they are okay with giving it to you. I have found this to be valuable when I have a simple question or important information to share and getting them on the phone is difficult.

6. Bring a list of current medications with you. Carry that list with you at all times. Even though I wear a Medic Alert bracelet, guess what the hospital prefers if you go through an ER? Yes, a simple (computer) printed list with all the medications you take and the allergies you have. This has helped me many times when I go to a new doctor or even a known one and they want me to write out my medications. I just hand them my list.

7. Get a Medic Alert bracelet, necklace or whatever. Hopefully you will never need it but it is good to have. This is especially important if you have diabetes.

8. Train your loved ones or close friends for what to do in an emergency for you in case you are rendered incapacitated: whom to call, where you need to go, your doctors' names and numbers.

While I have not mastered the art of advocacy at all times, I am predominantly happy with my dealings with my medical teams. I keep learning as I go with each new trial and tribulation CF or transplant throws at me. I feel that all of my experiences have helped me be good at communicating medical concerns, not just for myself but for my loved ones, too.

I now go with my mom to her doctor appointments, when possible. It turns out, she leaves a lot out when she talks to her doctors, but I am there to fill them in on what is really happening. And it is my pleasure to do so, as she doesn't want to deal. Funny how things have come full circle. I am fortunate to be able to help her advocate for the best care possible, even though she may not feel that way. She may not realize it, but she taught me well. ▲

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.*

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phage administration, Pseudomonas resistant to antibiotics are placed under pressure by the attacking phage, killing most of the bacteria. But a small portion can mutate and evade the phage attack, although the effort renders them again sensitive to antibiotics. At this point, antibiotics are administered to deplete the remaining bacteria. AB-PA01 is an investigational drug therapy targeting CF, as well as non-CF, Pseudomonas isolates and is designed to completely eradicate the infection. Currently in preclinical development, an early clinical study of AB-PA01 as a CF treatment using a nebulized formulation of the phage cocktail is anticipated for 2017.
http://tinyurl.com/jsulx9w

4-D Lung Scanner Able To Visualize Changes In Cystic Fibrosis

New four-dimensional computed tomography (4DCT) for lung scans (the 4DxV x-ray imaging technology) provides a state-of-the-art, noninvasive way of understanding regional lung motion and airflow in real-time within the breathing lungs. This enables highly detailed maps of both the patterns of lung motion and pulmonary function, with functional deficits detected through local differences in movement. The 4Dx scanner produces high-resolution images of lung-tissue motion and airflow, allowing investigators to view and measure abnormal lung function in specific areas before disease progresses and spreads. It offers images of the breathing lungs, making it possible to see what is really important — not what they look like — but how they work.
http://tinyurl.com/gtemwj7

Wireless Tech Augments Treatment For Cystic Fibrosis Patients

New technology from Hill-Rom Holdings is designed to more easily clear airways for patients with cystic fibrosis, and let clinicians know at a distance how patients are responding to treatment. The product, called the VisiVest System, uses high-frequency chest-wall oscillation, which helps to clear mucus from lung airways. It comes with wireless connectivity from Qualcomm that sends data to a portal from Razorfish that displays data trends to clinicians via a dashboard. The data also is available to patients so they can assist in treatment decisions with the goal of improved therapy adherence.
http://tinyurl.com/zuqge74

AND

http://tinyurl.com/hqudm9u

Mini-guts Predict Cystic Fibrosis Patients' Response To Therapy

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NOTES FROM CFRI'S 29TH NATIONAL CYSTIC FIBROSIS FAMILY EDUCATION CONFERENCE

Riding The CF Wave

By Laura Mentch

Among the shells, sand and surfboards decorating the Sofitel for the annual CFRI conference was the coming together of many people who care about those living with CF friends I've known for several years, those I've met online, family members, sponsors, researchers and healthcare providers. The thorough attention to infection control allows people living with CF to fully participate in sharing and receiving good information and great support.

Throughout the conference we learned of progress in understanding many aspects of the CF experience, new knowledge and developments in CF care and this refrain from many: **Participate in Clinical Studies.** This year there are more opportunities than ever before to further our understanding of CF. Patients are the key to the advancing knowledge and available treatments, the future is in our hands in partnership with researchers.

Alexandra L. Quittner Ph.D., in her presentation, "There Is No Health Without Mental Health," brought the results of her TIDES study showing a higher prevalence of anxiety and depression in people with CF and their caregivers. Depression in these groups was shown to decrease adherence to treatments and care. The Cystic Fibrosis Foundation, recognizing the need for mental health support, has initiated a pilot program placing mental health coordinators in care centers and providing education about depression and anxiety. Coming soon to a clinic near you!

Participate in Clinic Studies! Dr. Quittner is recruiting for her new study on resilience factors in adolescents and adults. Interested in helping? Contact ROSES at Miami.edu, 305-284-2097.

Putting research into action, CFRI (Cystic Fibrosis Research, Inc.) offers support for patients and families through these programs: Caregivers' (online) Support Group, Mothers' Retreat, Summer Retreat, Financial Support for Individual Counseling and Mindfulness-based Stress Reduction (online). Information about these opportunities can be found at http://www.cfri.org/support.shtml and http://www.cfri.org/mbsr.shtml or by calling CFRI at 855-237-4669 (toll free).

Raksha Jain, M.D., in her presentation, "Sex Hormones and Their Impact on Gender Disparities in CF Health," shared her quest to unwrap the disadvantage of being female with CF. Dr. Jain has looked at size of lungs/airways, adherence to therapies, co-morbidities (e.g., diabetes and nutritional status), physical activity and sex hormones. Though estrogen is believed to be the greatest factor, Dr. Jain's research will bring us further toward the truth. I learned that:

- FEV_1 decreases during the menstrual period,
- estrogen influences the change in pseudomonas from non-mucoid to mucoid,
- progesterone inhibits cilia activity in the airways and
- neutrophils are over-activated in the presence of estrogen.

More from my notes:

- Gallium, now being studied, kills resistant pseudomonas and is effective against biofilm.

PHOTO CRAIG BURLEIGH / BURLEIGHPHOTO.COM

LAURA MENTCH, USACFA DIRECTOR, AND RONNI WETMORE, RN, MS, COORDINATOR OF STANFORD ADULT CF CENTER, BOARD OF DIRECTORS OF CFRI.

• Colonoscopy screening is increasingly important in CF care with greater numbers of adults living with CF and concerns about colon cancer. Dr. Jordan Dunitz, from the University of Minnesota, recommends routine colonoscopy every three years for adults with CF and yearly colonoscopy screening for persons with CF who have had polyps. The care team in Minnesota has created a protocol for colonoscopy prep for persons with CF.

• Known mutations for CF now number ~ 2,000!

• Inflammation is a good thing and a bad thing: too much or too little inflammation leads to disease.

• Beth Sufian, J.D., reviewed the changes in Social Security Medical Criteria for CF and suggests we ask our care providers to keep good records that include the total time required for treatments.

• Gene editing to correct mutations in cystic fibrosis is on the horizon. We were given a book recommendation that may help us better understand the possibilities ahead: *The Gene: An Intimate History* by Siddhartha Mukherjee. The #1 *New York Times* bestseller from the Pulitzer Prize-winning author of *The Emperor of All Maladies*—a fascinating history of the gene and "a magisterial account of how human minds have laboriously, ingeniously picked apart what makes us tick."

The conference was bookended with personal stories, the heart and soul of CF.

On Friday night we heard from Ginny Dieruf of her family's tribute to their daughter in creating The Cody Dieruf Benefit Foundation for Cystic Fibrosis. In Ginny's talk, "From Adversity to Hope," we were introduced to Cody, her life and dreams and the foundation created in her memory that supports families in southern Montana with unmet expenses and opportunities that enhance their lives.

Emily Schaller in "Running Down a Dream: Emily's Mission to Rock CF," entertained and inspired us with the story of her path to increased health through running, cycling and physical activity while encouraging others with new sneakers and support. ▲

Laura is 63 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Meet A New Director Of USACFA

Greetings to my fellow readers of *CF Roundtable.* I am 24 years old, born and raised in Richmond, Virginia, where I currently reside. I was diagnosed with CF at 18 months of age. I am first generation Hungarian and proudly speak the Hungarian language. I plan to visit Hungary again in the next year. I have two older siblings and I am an aunt of three. I am the only one in my entire, extended family to have CF.

I graduated from Virginia Commonwealth University with a Bachelor's degree in Biology in December 2014. I currently work part time as a lab manager in an environmental microbiology laboratory at the university. Working with pseudomonas and other familiar CF microbes in the environmental setting is fascinating as these organisms, which unfortunately live in our lungs, are ubiquitous throughout the environment.

Within the past few years, I have become increasingly interested in being involved in the CF community by contributing as well as hearing others' ideas, tips and personal stories about living with CF. I have a very good doctor-to-patient relationship with my adult CF care team in Richmond, and I have been followed by the Duke Transplant center for the past few years, as I have lung function in the 30 percent range. Through their help, along with my very supportive family, I am fortunate to have accomplished as much as I have.

ELLA BALASA

I want to help educate and provide support to others who face the same challenges in fighting this disease. In addition to my daily health maintenance routine, I love to spend time outdoors in the summer months at the James River in Richmond or go on day beach trips to Virginia Beach. I also travel as much as I can and hopefully my next stop will be around Eastern Europe. I look forward to contributing to *CF Roundtable* and being a part of this great organization. ▲

Applications Due November 30, 2016, For USACFA Spring 2017 Scholarships

The United States Adult Cystic Fibrosis Association (USACFA) is excited to offer the Lauren Melissa Kelly Scholarship awards for Spring semester 2017. The scholarships will range from $1,500 to $2,500 and will be awarded to adults who have cystic fibrosis and are pursuing associate or bachelor degrees. Read on to learn more about Lauren Melissa Kelly and see below for scholarship criteria and requirements.

***Lauren Melissa Kelly** was an extraordinary woman**.** Laughing, gregarious, spontaneous, fun, talkative, driven, thoughtful, smart, kind and loving – all descriptive terms for Lauren, who lost her battle with CF late in her senior year at the University of Georgia.

In 1992 Lauren was chosen as one of ten Senior Leaders at the University of Georgia. She had distinguished herself as a member of the Golden Key Honor Society, Mortar Board, Phi Upsilon Omicron, Gamma Beta Phi, the Tate Society and Delta Gamma sorority.

Lauren felt the most significant opportunities of her college career included participation in the reconstruction and formation of organizations which will serve the university in the future. She acted as one of the refounding members of the Phi Kappa Literary Society and was significant in the metamorphosis of the Z Club into the William Tate Society. Her other activities included Greeks Advocating Mature Management of Alcohol (GAMMA) in which she served as Secretary and Special Events Chair. She was also a member of the Women's Glee Club for more than two years.

In recognition of her academic performance, Lauren's degree of Bachelor of Science in Family and Consumer Sciences was awarded posthumously. At the time of her death, Lauren was engaged to be married and living off campus in an apartment. She lived life to the fullest!

IN MEMORY OF
LAUREN
MELISSA KELLY

Walt Disney said, "Don't cry because it's over, smile because it happened. It's not the days in life you remember, it's the moments." As Lauren's mother stated, "I smile because she happened to me. Now, I want you to smile because she has happened to you."

**Lauren's bio was provided by her mother, who provided the scholarship grant in her memory.*

Criteria:

- The individual must be a United States Citizen and over the age of 18.
- The individual must have a positive diagnosis with cystic fibrosis.
- The individual must be pursuing a bachelor or associate degree.
- The individual should not be an immediate family member of someone on the USACFA board.

Requirements to include in the application:

- The USACFA Scholarship application form (includes the three essays)
- A copy of an official transcript from high school (and current college, if you have completed coursework)
- A copy of proof of enrollment for the Fall 2016 or Spring 2017 semester
- A resume
- A letter from the doctor/clinic on letterhead confirming cystic fibrosis diagnosis
- A headshot photo

Deadlines:

ALL requirements must be sent in PDF or other appropriate digital format to scholarships@usacfa.org by **Wednesday, November 30, 2016, at 11:59 p.m. EST.**

- Application Form and Instructions
- Download our http://www.cfroundtable.com/wp-content/uploads/United-States-Adult-Cystic-Fibrosis-Association-Scholarship.pdf form to your computer.
- Open and fill out the application form using your computer.
- Once the form is completed, please save the file.
- Make sure all other requirements (from above) are completed in an appropriate digital format and attach them in a single e-mail sent to scholarships@usacfa.org.

PHOTO BY STEPHEN BOYER

Hand in Hand

Surrounded by sickness
Together we stand
Kaeti and Allison
Hand in hand
One without the other
We're not as strong
Life doesn't make sense
Everything seems wrong
But with each other
At our sides
We are in control
And take great strides
To living life however
We desire
A thought to CF
But it's a mere flat tire
Because in the long journey
It's a small anecdote
But our lives are our stories
CF is the sidenote

-A. Best, 2004

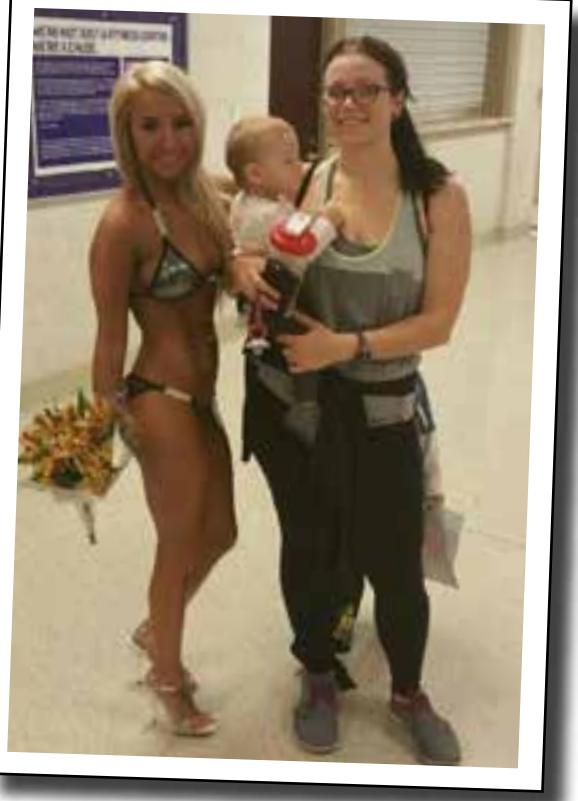
"Through the Looking Glass: Images of Adults with Cystic Fibrosis" and "Caregiver Stories" are projects of Breathing Room, a non-profit organization. Breathing Room hosts these and other projects to facilitate open and candid communication in the CF community, supports the development of a community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

FROM OUR FAMILY PHOTO ALBUM...

ISABEL STENZEL BYRNES WITH THE USACFA JACOBY ANGEL AWARD AND BETH SUFIAN WITH THE CFRI PARTNERS FOR LIFE AWARD GIVEN IN MEMORY OF ANABEL STENZEL.

APRIL HANSEN AND CRIS DOPHER CELEBRATED THEIR THOUSANDTH DAY POST-TRANSPLANT ON SEPTEMBER 5, 2016. THEY WERE BOTH TRANSPLANTED AT DUKE ON THE SAME DAY, DECEMBER 13, 2013.

TOP L TO R: CODY, NOAH, COLE, AND JORDON; BOTTOM L TO R: TERESA, SAM, GARETTE, AND PRESTON LANGMEAD.



APRIL SCHOEN AND FRIEND, MELISSA, AT "MY DREAM BODIES CLASSIC," IN TOLEDO, OHIO, 2015.

ELLA BALASA IN THE DOMINICAN RUPUBLIC, 2016.

LINDA STRATTON.

VICTOR ROGGLI, M.D., AS ELVIS PRESLEY.

FAMILY PHOTO ALBUM CONTINUED...

JEANIE AND JOHN HANLEY IN LA JOLLA, CA, ON JUNE 28, 2016 CELEBRATING THEIR ANNIVERSARY.

ANDREA EISENMAN, STEVE DOWNEY AND THEIR DOGS AT THE LOOKOUT IN MOUNT ROYAL PARK, MONTRÉAL, CANADA.

TODD VOWELS AND HIS NEPHEWS, LUKE AND GRIFFIN BARTON, SNOW-MOBILING AT MONARCH PASS IN COLORADO.

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CF Roundtable

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IN THE SPOTLIGHT

With Victor Roggli, M.D.

By Andrea Eisenman and Jeanie Hanley

You do not see this every day in the world of CF: someone who just turned 65 and is homozygous for delta F 508! Victor is a pathologist with an analytical mind, doing well in his career. He is a singer, a father and happily married – not necessarily in that order. He is outliving the odds and offers some explanations as to why – from keeping bacteria on their toes to why men and women differ in health when it comes to having CF.

Please welcome our newest star. Spotlight, please!

Diagnosed when and why?

When I was seven years old, I came in from working in the garden on a hot July day. My mom noticed that I was white as a ghost and had fine crystals (a primitive sweat test) on my shoulders. In the next two weeks I was diagnosed with mumps, measles and double pneumonia by my hometown doctor. From that day forward I had a chronic cough that wouldn't go away and I failed to gain weight in spite of a ravenous appetite.

At age ten, my parents took me to an allergist in Chattanooga, TN, who sent a letter to my hometown doctor recommending a sweat test. It was filed in my chart but ignored. Finally, in desperation, my parents took me to Vanderbilt in Nashville, TN, where a medical sweat test was administered and I was officially diagnosed with CF, in 1963, at age 12.

Any siblings with CF?

I have two sisters and one brother, none of whom had CF. However, four years before I was born, my mom had a baby boy who died at three months. The death certificate said "whooping cough." In retrospect, we suspect that he had CF.

Married? Kids?

I was married at age 19 (12 days shy of my 20th birthday) and have one daughter (38) by artificial insemination. I am now married to my second wife, Linda; we recently celebrated our 25th wedding anniversary.

VICTOR ROGGLI, M.D., WITH HIS NEW MEDICARE CARD.

How has CF affected your relationships with the opposite sex?

Because of my poor nutrition, I lagged behind my peers developmentally, which caused me considerable anguish and made me feel inferior (who would want a scrawny little kid with a chronic cough?). Indeed, from the time I was diagnosed at age 12 until I turned 14, I doubled my weight from 70 to 140 pounds! I also felt my intelligence was a liability rather than an asset in dealing with the opposite sex. Frankly, I was a mess emotionally for many years, which my darling wife Linda helped me to work through.

What health issues do you have?

As I get older, more things seem to be going wrong. I have been hospitalized on a couple of occasions (age 21 and age 32) for coughing up blood and on a couple of occasions (age 41 and age 62) for pneumonia. I have also had a couple of hernia repairs, which along with GE reflux is probably a consequence of chronic coughing. In recent years I have had a PICC line with two to three weeks of IV antibiotics about once per year. For the past 13 years I have also been dealing with cardiac arrhythmias with a fast heart rate, culminating in an ablation procedure via cardiac catheterization this past March. Also, for the past year, I have been dealing with gastroparesis (feeling of being full after eating a relatively small amount), a frustrating turn of events for someone who has always been able to eat as much as I wanted of whatever I wanted. Gastroparesis and several attacks of gout have put a damper on that. Seems like it is always something!

Do you exercise? If so, what?

I used to get up every morning and shoot baskets in my driveway for 15 minutes. However, since I have been dealing with the fast heart rate issues, it has been difficult to exercise on a regular basis. With the approval of my cardiologist and pulmonologist, I hope to get back to a regular exercise program in the near future.

What interested you in becoming a doctor?

From the time I was young, I was interested in medical research. My col-

lege advisor suggested that I go to medical school. So I graduated from Baylor College of Medicine in Houston, TX. After medical school, I trained to be a pathologist (with a specialization in lung disease. Surprise! Surprise!). I am currently Professor of Pathology at Duke University Medical Center in Durham, NC, where I have been for 36 years. I am still working full time with no plans to retire.

I chose pathology because it was a great specialty to perform research (I have published over 200 articles in medical journals). Also, I was afraid I would work myself to death in internal medicine and surgeons got up too darned early in the morning (I am not an early morning person!).

How did it feel to receive your Medicare card?

Definitely mixed feelings! On the one hand, I felt old. I remember as a child thinking that people who were 65 were really old. On the other hand, I was 65, with CF, and still alive! It reminded me of a quote from George Bernard Shaw who was asked on his 95th birthday how it felt to be 95. His answer: "Pretty good when you consider the alternative!"

What do you attribute to your longevity with CF?

It's a very complex issue. The support system that I have from my loving wife Linda is, of course, key. I have always been meticulous about taking all of my prescribed medications and rarely miss a dose. I welcome all opportunities to try promising new treatments. I also have a very strong immune system, which I suspect suppresses my nonspecific immune response to the multiple types of bacteria that colonize my lower respiratory tract. This nonspecific reaction (mediated by white blood cells or neutrophils) is responsible for much of the lung injury in CF. There is some research to support this theory; steroids (which suppress your specific immune system) are not effective for CF patients. Non-steroidal anti-inflammatory drugs (like ibuprofen) which suppress your nonspecific immune reaction actually do help. Finally, one of my physicians suspects that my love of singing has produced a strong diaphragm which helps with mucus clearance.

How did you get into singing?

I loved singing along with the radio as a teenager. When I was 21, I got my first guitar and taught myself how to play guitar. Then I started making recordings of myself playing and singing. When karaoke became popular in the 1990s, I fell for it hook, line and sinker. I have a fairly sophisticated home system with more than 3,000 karaoke songs and I have recorded more than 300 songs. I regularly host karaoke parties at my house for medical students and trainees.

Elvis has always been one of my favorite artists and I probably do a better impersonation of him than any other artist. My wife threw big parties for my 50th and 60th birthdays, and I did several Elvis songs at each one. For my 60th, she sent out a "save the date" brochure with the picture of me dressed as Elvis.

Do you use supplemental oxygen?

In early 2015 I discovered that I was desaturating with moderate activity. I had already been diagnosed with sleep apnea several years before. So last year I began using oxygen on a routine basis: 3 L continuous flow at night and 6 L intermittent flow with activity (such as walking 100 steps or more or climbing steps).

Describe your use of the "Trilogy System" and how it has helped you.

At about the same time as the desaturation discovery, my physician determined that I had mild CO_2 retention, which goes along with sleep apnea. Since I have been using the Trilogy System (a sophisticated CPAP/BiPAP machine) routinely, my CO_2 has gone down nicely.

Maintaining my oxygen saturation and keeping the CO_2 down helps to protect my heart, which takes a lot of wear and tear with the stresses it receives from my less-than-adequate lungs.

Discuss your dandruff shampoo theory in treating CF.

Years ago, there was a dandruff commercial in which one fellow claims that he was able to control his dandruff by switching shampoos. Later, a dermatologist affirmed that the commercial was true, simply because each brand of shampoo has a different active ingredient. The fungus that causes dandruff is thwarted by one shampoo for a while, but eventually becomes immune to its effects. Switching brands exposes the fungus to a new active ingredient.

I think the same logic applies to CF. The bacteria that colonize my lungs get accustomed to whatever treatment I send their way and they build up a resistance to that medication. By continually trying new treatment approaches, I have managed to keep my Pseudomonas "free-loaders" off balance. I believe that is one of the reasons in the long run that I have done so well.

What is your theory about men living longer with CF than women?

I think it has to do with anatomy and the physiology of coughing. The right upper lobe is a problem area for many patients with CF. The right lower lobe bronchus comes straight off the windpipe with very little angle, whereas the right upper lobe bronchus is at a right angle from the windpipe (and has a smaller opening). A cough can clear the mucus more readily from the lower lobe with the straight shot up the windpipe, whereas it is much less effective for that right upper lobe. Women are smaller and have smaller lungs with smaller bronchial openings, so they

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ROGGLI continued from page 25

have more trouble clearing mucus.

What do you think will give others living with CF hope?

I have witnessed remarkable changes in treatment in the decades since I was diagnosed. The multidisciplinary approach to medicine, centers of excellence for the treatment of CF and continuing improvement in lung transplant technology and gene therapy bode well for continuing improvement in CF prognosis. Although we are not quite "there" yet, the promises of gene therapy are tantalizing. For those of us for whom the ravages of years with CF have rendered our lungs impervious to the effects of gene therapy, the improvements in lung transplantation have been amazing. And stem cell research holds the promise of taking your own stem cell, removing the bad CF gene and replacing it with a good one, and then growing a new pair of lungs in the test tube. Voila! No need for antirejection medications and the infectious complications that go along with them. Those of us who have made it to the mountaintop, so to speak, by making it to 60, are paving the way so others can follow.

Any advice to offer to others with CF?

Follow your dreams. Make sure you are at the wheel and that CF is in the back seat. It is not realistic (nor very smart) to totally disregard CF, but you can certainly treat it as a nuisance, an inconvenient truth, so to speak, rather than letting it dominate or rule your life. ▲

Andrea Eisenman is 51 and has CF. She is a Director of USACFA and is the Webmaster and Executive Editor of CF Roundtable. *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 15

Mini-guts grown in the lab using cystic fibrosis (CF) patients' cells can help pinpoint those who are most likely to benefit from new drugs, according to a new study. The findings pave the way to exploiting so-called organoids as a tool for screening drugs and personalizing treatment for patients with CF and other genetic diseases. CFTR-targeting therapies have recently been approved to treat CF, but identifying the subset of patients most likely to respond to these drugs remains challenging, time-consuming and costly. To better predict individuals' clinical response, the researchers grew gut organoids, three-dimensional adult stem cell cultures that resemble the gut, using rectal biopsies from 71 CF patients who carried a wide range of CFTR mutations. They found that the mini-guts' responses to CFTR-targeting drugs in vitro closely correlated with published data from clinical trials of the same drugs. Based on these findings, they selected two individuals with rare CFTR mutations whose rectal organoids responded strongly to the treatment. When given the drug, both individuals showed fewer disease symptoms and improved lung function. The results support the potential to use patient-derived organoids to tailor treatments for individuals.
http://tinyurl.com/jchaw6j

Long-term Tiotropium Bromide Therapy In CF Patients Linked To Improvements In Lung Function Decline

Adult patients with cystic fibrosis (CF) show improvements in lung function decline when treated with a high dosage of tiotropium bromide taken over a long term. A team of researchers tested the effects of tiotropium bromide when administered for 24 months, and at a high dosage of 18 micrograms. Patients treated for 24 months with tiotropium bromide had a significantly slower decline of mean annual change of FEV_1 compared to placebo controls. In patients with a FEV_1 of 70 percent predicted or greater, long-term tiotropium bromide treatment was associated with greater improvements in annual lung function decline. No effect of the treatment in the mean annual change of exacerbations in adult cystic fibrosis patients was seen.
http://tinyurl.com/z4j7f2z

Inhaled Alkaline Buffer Could Help Cystic Fibrosis Patients Clear Lungs Of Bacteria

Increasing the pH balance of the acidic lung surface liquid found in cystic fibrosis (CF) patients, using an inhaled alkaline buffer, helps to kill bacteria in the airways. The study proposed that the buffer, called trometh-amine or Tham, could be used as a treatment option for managing airway symptoms in CF patients. The pH of the thin film of liquid produced in the airways is a crucial feature largely determining its function. In addition to preventing the lungs from collapsing when we exhale, the liquid contains antimicrobial peptides — our body's own antibiotics — that protect the lungs from invading bacteria and other types of infections. Starting by collecting sputum samples from patients, the team tested the effect of a sodium bicarbonate alkaline buffer to restore pH. Results showed the sodium bicarbonate both restored pH and allowed more effective killing of the bacterium Staphylococcus aureus (S. aureus) — one of the first bacterial species to colonize CF lungs. The team tested Tham and found this buffer improved the activity of two antimicrobial peptides called hBD-3 and LL-37. Since CF patients are often treated with high concentration salt solutions to ease the transport of mucus from the airways, researchers also tested if the inhaled buffer would be effective in combination with salt, and found that the presence of the salt solution lowered the buffer's bacterial killing capacity by only a very small degree. Researchers concluded that the inhaled Tham buffer, either alone or in combination with a hypertonic saline solution, might be of therapeutic benefit for CF patients.
http://tinyurl.com/h76zdg6

Breathing In A Cure: Inhalable Ibuprofen On The Horizon

Research has found that ibuprofen, when taken at high doses, helps slow the progression of lung function decline in people with cystic fibrosis. The downside is that ibuprofen doses that high, when taken routinely, can result in gastrointestinal (GI) bleeding and acute kidney injury. Researchers feel that nanoparticle ibuprofen delivered by aerosol to the lungs would be a fantastic therapeutic. The inhaled ibuprofen would work in conjunction with the antibiotics the patient is already being given for the underlying infection as it was determined that not only does ibuprofen act as an antimicrobial itself, it is also synergistic with the antibiotics already given to patients.
http://tinyurl.com/jpbbfnb
AND
http://tinyurl.com/hg3woe9

PUR1900 For CF Patient Lung Infections Granted Orphan Drug Status

Pulmatrix's inhaled PUR1900 for treating pulmonary fungal infections in patients with cystic fibrosis (CF) has received orphan drug status by the U.S. Food and Drug Administration. The designation was granted based on a likely theory that PUR1900 may be clinically superior to the same drug that is already approved for the same infection. Recent studies demonstrated that itra-

Continued on page 31

ACTIVE FOR LIFE

Sneak It In

By Aimee Lecointre

I don't know about you guys but it's not every day that I want or am motivated to hike mountains, hit the trails running, lift weights, get on my yoga mat or walk on the treadmill. There are times I don't even feel like walking to the bathroom! Some days I just don't feel like setting time aside to work out, others I just don't have the time and others still I just feel too awful.

The thing is, being active and exercising are among the most important things to help me maintain my health, so I try my best to do what I can. I believe every little bit adds up. Anything we can do to help our bodies, especially our lungs, is worth it.

Here are some tips to sneak in some activity to get moving, get your heart rate up, get coughing and maybe even break a sweat on those days you just don't have time or don't feel like doing much.

1. Don't always choose the closest parking spot.

When I feel up to it I try to pick a parking spot farther away from the store so I have to get in some extra steps.

Take the stairs.

2. Use the stairs when you are feeling up to it is a great way to sneak in some extra steps and even cardio. On days when I'm not feeling 100 percent, and there are a lot of floors I need to get up, I will take the stairs part of the way, then the elevator the rest.

3. Walk the dog.

If you have a dog don't get caught in the rut of just letting it out back to do its business. I try to take my dogs on a walk at least once a day, weather and lungs permitting. Your lungs will benefit and your dog will be so happy! And if you don't have a dog try to get out for a walk, even if it's just to the end of the driveway and back.

4. Every hour on the hour.

When I'm at work or even at home some days I will try to get in one minute of exercise every hour on the hour. Some days it's over a period of four hours, others it's eight. Try things like jumping jacks, lunges, squats, push-ups, burpees (if you're super motivated), tricep dips on a chair, calf raises, butt kickers etc. Sometimes even just stretching here and there feels great.

5. Stretch in bed.

Some days are better than others and some days you just don't want, or can't, get out of bed. Doing some simple stretches if you are able is great, especially if you can stretch out your chest area. Here's a great stretch for the chest: sitting up in bed, reach your arms above your head and grasp just below your right wrist with your left hand, then gently pull the right arm over to the left and let the body follow. You should feel a nice big stretch in the right side of the body, breathe into all that space. Hold for a few breaths, then go back to center. Repeat on the other side.

> *"Being active and exercising are among the most important things to help me maintain my health, so I try my best to do what I can."*

AIMEE LECOINTRE

6. ACBT - Active Cycle Breathing Technique.

I don't know about you, but some days after a round or two of ACBT I feel like I got in a decent workout. Even if your energy is low or if you aren't physically able to do much activity. Keeping the lungs clear is key, not that I need to tell you guys that!

7. Stand at work.

If you work all day at a desk, get a standing desk or create one with some books or a sturdy box. Even just standing can be better than sitting all day. Unless I am writing a new article, blog post or creating content for my website, most of my work can be done while standing. I just seem to write better while sitting in a comfy chair! Using a yoga ball instead of a desk chair is another great option. I used a yoga ball as a chair at my last office job and

noticed an improvement in my core strength and my posture.

8. Sneak it in during TV time.

Try some of the exercises from tip number four and sneak in some exercise during commercial breaks.

9. Dance party!

Personally this is my favorite tip. Have an impromptu dance party. Turn on some of your favorite music and just dance it out like no one's watching! I don't know about you, but it doesn't take much dancing for my heart rate to go up and my lungs to get working!

Remember:

- Every little bit counts.
- Don't compare your health or level of activity to others. You do you!
- Try your best. Some days that's all any of us can do.
- Listen to your body. If you need to rest, then rest. Don't push yourself so much that you end up feeling worse. Some days, just lying in bed is what we need or all we are capable of in that moment.

I would love to hear from all of you your tips for sneaking in exercise or activity! Feel free to e-mail me with your ideas. ▲

Aimee is 31 and has CF. She lives in Utah with her husband, two pugs and a cat. She is a registered yoga teacher and nutritional therapy practitioner. She can usually be found hiking in the mountains, spending time with family or cooking up something delicious in the kitchen. You may contact her at: alecointre@usacfa.org.

POETRY CORNER

Struggling To Breathe

By Linda Stratton

In the twilight of the morning I lay listening to the percussion of my heart and lungs.
With cystic fibrosis, lungs fill with congestion, strangling every effort to breathe.
Unable to lie down in an attempt to sleep, pillows are stacked behind my back.
Breath comes in short bursts, with sound like bellows compressed—a raspy cough repeatedly racking my body.

Struggling to breathe, I fight the fear and panic lying just beneath the surface, threatening each moment to betray me.
At one time anger was my only emotion, blaming God for this disease within.
Now, I take comfort in Him as my struggle for breath—for life, is at hand.

Even as I fight to breathe, I feel God is near.
He gives me strength when my body is weak—courage when my hidden desire is to succumb.
I'm clothed with peace as the reality of my future is set before me.
In the moments between confidence and fear, I'm thankfully blessed with faith enough to overcome.
This state of mind brings freedom—freedom from worldly concerns and mere afflictions of the body.
As I trust in Him, each breath comes more easily; He nurtures my body as well as my soul.

Linda is 62 and has CF. She has a heart for the elderly and enjoys living with and caring for her 89-year-old father, James, in Denver, Colorado. Other interests include church volunteer work and crocheting.

SEARCHING FOR THE CURE

On Attending My First CF Conference And The Next Generation Of CF Therapies

By Reid D'Amico

This past July, I had the opportunity to attend my first CFRI conference. Ideas swarmed my head as to what the conference would be like. I had never attended a CF conference before, so I had little grounding on which to base my initial guesses. Would the conference be advocate-driven? Would there be groundbreaking science talks? Would this be a networking event? In short, all of these answers turned out to be yes. The CFRI conference was a perfect match of science, advocacy and personal stories with the common thread of hope. We all have an optimistic view of the future of CF care, but there was something special about meeting the minds behind the future of our health.

Leading up to the conference, I remember being apprehensive about CF safety guidelines. Due to cross-infection issues seen in CF, I knew this would pose a unique and interesting challenge for this conference. After filling out paperwork, retrieving doctor's notes and reading the guidelines, I quickly became aware that CFRI takes infection control very seriously.

Upon arriving at the conference, the first thing I noticed was the arsenal of hand sanitizers at every corner. I loved this! If I had my way, every conference and event would have hand sanitizer everywhere. After starting off on this good foot, I was even more pleased to see the other guidelines in effect. Those of us with CF were not allowed to dish up our own plates in the food line and all drinks were served for us. Rare chronic illness aside, being wined and dined always is nice. So for anyone who is apprehensive about attending CF conferences, be sure to read the guidelines and know that they are incredibly diligent in preventing the spread of bacteria, viruses and fungi.

With my mind at ease about catching some nasty infection, I was able to delve into what the conference had to offer. The CF vendor fair was incredible. As someone who attends academic conferences often, it was nice to see what other companies and organizations could offer *me* to make *my* CF care a little better. I was able to find a new pharmacy, see the new cutting-edge technology and make some friends in the CF community. We're familiar with how CF can be an isolating disease, so the fair offered a unique opportunity to meet the employees, advocates and scientists who dedicate their lives to our disease.

> *"What did excite me was the next generation of CFTR modulators, mucus hydrators and anti-infective treatments."*

REID D'AMICO

However, as a Biomedical Engineer, I was looking forward to the science talks the most. The talks largely comprised faculty and fellows at Stanford and the University of California, San Francisco (UCSF). The talks ranged from mental health research and gender disparities, to basic science seminars. All had the underlying tone of pushing for a more comprehensive understanding and treatment of all types of CF. As an engineer, I was in awe of the progress made in science to understand CF, and I was able to see where the mix of science and engineering could expedite and re-create the future of CF research.

From a clinical trials perspective, I felt an unwavering sense of excitement. The onslaught of new therapies entering the pipeline or ongoing clinical trials will inevitably change the face of CF care. Of course, CRISPR gene editing technology headlined the talk by discussing the potential to change the

underlying mutation in the DNA. However, as a medical researcher, I know that the hurdles of this technology will prevent it from entering the clinical trials sphere for many years. What did excite me was the next generation of CFTR modulators, mucus hydrators and anti-infective treatments.

In my own care regimen, I have always found that hypertonic saline has been my crutch. Few other medicines give me such a sense of relief, so hearing about the new airway surface liquid restoration left me optimistic for even better treatments. Curiously, I found myself most fascinated by a new type of antibiotic treatment: gallium. Many may remember the image of this metal melting in someone's hand from our chemistry classes, but we're starting to shed light on gallium's ability to combat biofilms in cystic fibrosis. Like hypertonic saline, I'm always amazed by how simple therapies can treat a disease as complicated as CF.

To me, the conference masterfully attracted the multifaceted front line of CF. Patients, parents, family members, scientists, doctors, advocates all gathered in one location to celebrate the successes and the future of CF. For that, I am immeasurably grateful to have had the opportunity to attend the CFRI conference. To anyone who attends a CF conference for the first time, I promise you will take away more than you expected—but really, the vendors give away wonderful table gifts and multivitamins, and I also won a pizza-making kit. It was the cherry on top of a great weekend. ▲

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

TILLMAN continued from page 27

conazole taken orally is effective in the treatment of allergic reactions in CF patients with fungal lung infections. High doses are needed to receive enough of the antifungal drug in the lungs through the bloodstream, but side effects including liver toxicity can be severe. This technology delivers the drug directly to the lungs, which reduces the risks of side effects and drug-drug interactions. PUR1900 is an inhaled anti-fungal that combines itraconazole with the company's iSPERSE technology that allows patients to inhale the drug easily. The iSPERSE technology aims to reduce the toxicity that results from high doses of itraconazole.

http://tinyurl.com/hkqy6u7

AND

http://tinyurl.com/zm6bqp7

CF Foundation Gives Alaxia $1.7M To Start Testing An Inhaled Antimicrobial Therapy In People

Alaxia SAS announced that it has received $1.7 million from the Cystic Fibrosis Foundation Therapeutics (CFFT) to support and advance a first clinical study of its inhaled antimicrobial drug candidate, ALX-009, as a therapy for people with cystic fibrosis (CF). The randomized, double-blind and placebo-controlled Phase 1 trial will evaluate the safety, tolerability and pharmacokinetics of single and multiple ascending doses of hypothiocyanite (a bacterial agent), bovine lactoferrin (an iron-binding glycoprotein derived from cows) – two compounds with antimicrobial properties – or their combination (ALX-009) in healthy male volunteers and CF patients. ALX-009 is designed to compensate for the defective lung defense system innate to CF by delivering two key endogenous antimicrobial substances directly to the lungs. The drug's efficacy is not altered by complex structures like biofilm or sputum, often found in the lungs of CF patients. In vitro tests have demonstrated the therapeutic potential of ALX-009 against a broad range of bacterial species and particularly against clinical isolates with natural or acquired multi-drug resistance. ALX-009 can potentially limit the emergence of resistance – and induction of cross-resistance – to available antibiotics, the company reported.

http://tinyurl.com/j822fys

Laurent Pharmaceuticals Receives U.S. $3M Development Award From Cystic Fibrosis Foundation Therapeutics

Laurent Pharmaceuticals, Inc., announced it has received a Therapeutics Development Award of up to $3 million from U.S.-based Cystic Fibrosis Foundation Therapeutics, Inc. The award will help support the Phase 2 clinical trial of the company's lead drug candidate LAU-7b in adult patients with cystic fibrosis (CF). LAU-7b, a once-a-day oral solid dosage form of fenretinide, works by correcting the defective metabolism of arachidonic acid (AA) and docosahexanoic acid (DHA), and modulates chronic inflammation via a pro-resolving mechanism supported by a strong rationale linked to the expression of the CF genetic defect. AA and DHA are two essential fatty acids playing a crucial role in maintaining an effective immune-inflammatory response. The CF gene defect causes exaggerated AA-mediated inflammation and low DHA-mediated resolution, leading to lung infection and local tissue destruction. Resolution of inflammation is a new therapeutic approach using the body's own ability to

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MILESTONES

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

ANNIVERSARIES

Wedding

Andrea Eisenman and Steve Downey
New York, NY
8 years on September 13, 2016

Jeanie and John Hanley
Manhattan Beach, CA
30 years on June 28, 2016

NEW BEGINNINGS

Engaged

April Schoen and Kyle Purkey
Maumee, OH
Engaged June 28, 2016

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modulate inflammatory responses, thus addressing inflammation without inducing immunosuppression. LAU-7b was recently tested in adult patients with CF in a dose-ascending Phase 1b study, showing a good safety and tolerability. A Phase 2 clinical trial in adult patients with CF is currently in preparation.
http://tinyurl.com/jnaqqgr

Cystic Fibrosis: Lung Microbial Environments Change With Lung Function Decline

A recent study confirmed that different structure and composition of airway microbiota (symbiotic helpful and harmful microrganisms) exist in cystic fibrosis (CF) patients with stable or declining lung function; and shed new light on the fact that microbiota in stable lungs provides higher resilience to bacteria than lungs in severe function decline. Future research could reveal how interactions between microbial communities change upon immune response or treatment, and unlock potential development of microbiome-based personalized medicine for diseases like CF. The microbial community in CF patients' airways harbors a vast array of bacterial species not previously identified and suspected to be involved in the disease progression. For the recent study, researchers aimed to characterize extensively the microbial species present in patients' airways and how they impact disease progression. Researchers collected sputum samples from CF patients and identified the microbe content by species through sequencing. In order to focus on how the lung composition changes following severe decline, they compared the airway microbiota of CF patients with a substantial decline in lung function to microbiota of patients with stable lung function. Results showed 22 distinct bacterial genera in CF patients' airways; mostly Pseudomonas and Staphylococcus. Staphylococcus aureus was more abundant in stable lung function patients than in those whose lung function was already in decline. Notably, researchers identified several members of Prevotella genus, which are considered emerging CF pathogens. The presence of multiple bacterial genomes (denoting the polymicrobial nature of CF infection) was also highlighted by the presence of other minor genera, including members of the Alcaligenaceae and Carnobacteriaceae families. These results showed a different structure and composition of airway microbiota in CF airways of patients with stable and declining lung function. These differences suggest that the microbiota of stable lungs carry higher resilience, when compared to severe lung function decline patients – which impacts the aggressiveness of pathogens such as Pseudomonas.
http://tinyurl.com/ht7xuhf

Breathing In Bile May Promote Lung Disease And Antibiotic Resistance In CF

When patients with cystic fibrosis (CF) breathe in small amounts of bile, some bacterial species switch to a chronic infection mode, aiding both bacterial colonization of the airways and the development of antibiotic resistance. The study delves into a little explored research area, suggesting that efforts to prevent gallbladder fluid from reaching the lungs could help slow disease progression in CF. Gastroesophageal reflux disease in CF patients causes bile to rise, ending up in the airways. Studies have shown that up to 80 percent of patients aspirate bile. Since Pseudomonas aeruginosa is the microbe responsible for the vast majority of disease complications among CF patients, researchers decid-

Benefactors

BRONZE

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Helen Kleine (In memory of Jerome Feld)
Mr. & Mrs. Richard Millikan (In memory of Nahara Mau)
Kris & Kim Newport (In celebration of our daughter Jessica's 30th birthday)
Dr. Dan Seilheimer
Beverly & Dave Sufian (In honor of Beth & Sandy Sufian)
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$5,000 - $9,999 SUSTAINING PARTNERS
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ed to study how bile can affect the virulence of this bacterial species, as well as its response to antibiotics. An analysis of the bacterial transcriptome showed that bile triggered a host of changes in bacterial gene activity. Researchers realized that many of the altered genes were involved in activating a chronic infection state, helping bacteria to grow into biofilm. Once the biofilm is formed, they also became more resistant to antibiotic treatment with three commonly used drugs: colistin, polymyxin B and erythromycin. The team concluded that the data suggest that bile acid signaling is a leading trigger for the development of chronic phenotypes underlying the pathophysiology of chronic respiratory disease.
http://tinyurl.com/h6tqf4z

TREATMENTS

Long-term effects of azithromycin in patients with cystic fibrosis. Clémentine Samson, Aline Tamalet, Hoang Vu Thien, Jessica Taytard, Caroline Perisson, Nadia Nathan, Annick Clement, Pierre-Yves Boelle, Harriet Corvol. Respiratory Medicine. August 2016Volume 117, Pages 1–6

Low-dose azithromycin has beneficial effects on severity of the lung disease in cystic fibrosis (CF) patients for a period of 6 - 12 months after initiation of the treatment. Although its impact in the longer term is uncertain, this treatment is frequently used chronically. The aim of this retrospective study was to investigate the effects of low-dose azithromycin treatment on the progression of CF lung disease in patients treated for more than 12 months. No clinical benefits of low-dose azithromycin were present after one year of treatment in young CF patients. Selection for macrolide-resistant strains of bacteria occurred, which should lead to a reconsideration of the duration of azithromycin treatment in CF.
http://tinyurl.com/zx5n35t

Ursodeoxycholic acid treatment is associated with improvement of liver stiffness in cystic fibrosis patients. van der Feen C, van der Doef HP, van der Ent CK, Houwen RH. J Cyst Fibros. 2016 Jul 29. [Epub ahead of print]

Liver stiffness measurements to evaluate the effect of Ursodeoxycholic acid (UDCA) on the progression of CF liver disease were utilized. The researchers found that UDCA reduced liver stiffness in patients with well-defined, mild liver disease.
http://tinyurl.com/hj3vvoe

Safety and efficacy of prolonged levofloxacin inhalation solution (APT-1026) treatment for cystic fibrosis and chronic Pseudomonas aeruginosa airway infection. Elborn JS1, Flume PA2, Cohen F3, Loutit J4, VanDevanter DR5. J Cyst Fibros. 2016 Feb 26. [Epub ahead of print]

Levofloxacin inhalation solution (LIS) is the first aerosolized fluoroquinolone licensed for treatment of patients with cystic fibrosis (CF) and chronic Pseudomonas aeruginosa lung infection. This study evaluated the safety and efficacy of extended LIS treatment and found that extended treatment with LIS in 88 patients was well tolerated with no new safety signals and evidence of positive effects on FEV_1 and quality of life.
http://tinyurl.com/nukgsau

Timing of dornase alfa inhalation for cystic fibrosis. Cochrane Database of Systematic Reviews. 2016, 7: CD007923

This review was done to determine the effect of timing of dornase alfa inhalation on measures of clinical efficacy in people with cystic fibrosis (CF) (in rela-

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tion to airway clearance techniques or time of day). The authors concluded that the current evidence derived from a small number of participants does not indicate that inhalation of dornase alfa after airway clearance techniques is more or less effective than the traditional recommendation to inhale nebulised dornase alfa 30 minutes prior to airway clearance techniques, for most outcomes. In the absence of strong evidence to indicate that one timing regimen is better than another, the timing of dornase alpha inhalation can be largely based on pragmatic reasons or individual preference with respect to the time of airway clearance and time of day. Further research is warranted.

http://tinyurl.com/hug7aqy

Killing effect of nanoencapsulated colistin sulfate on Pseudomonas aeruginosa from cystic fibrosis patients. E. Sans-Serramitjana, E. Fusté, B. Martínez-Garriga, A. Merlos, M. Pastor, J.L. Pedraz, A. Esquisabel, D. Bachiller, T. Vinuesa, M. Viñas. Journal of Cystic Fibrosis. September 2016 Volume 15, Issue 5, Pages 611–618

The aim of this study was to explore antimicrobial activity of nanoencapsulated colistin (SLN-NLC) versus free colistin against P. aeruginosa clinical isolates from CF patients and to investigate their efficacy in biofilm eradication. The researchers found that colistin nanoparticles had the same antimicrobial activity as free drug against planktonic bacteria. However, nanoencapsulated colistin was much more efficient in the eradication of biofilms than free colistin. Thus, these formulations have to be considered as a good alternative therapeutic option to treat P. aeruginosa infections.

http://tinyurl.com/ze7te5k

Inhaled Mannitol as a Therapeutic Medication. Daviskas, Evangelia PhD, MBiomedE; Anderson, Sandra D. DSc, PhD. Clinical Pulmonary Medicine: September 2016 - Volume 23 - Issue 5 - p 197–202

Inhaled dry powder mannitol, being an osmotic agent, improves hydration at the airway surface, which in turn improves the physical properties of mucus and facilitates its clearance. These changes once sustained have been shown to improve or at least sustain lung function, reduce mucus plugging and biofilm formation, reduce the incidence of exacerbations, increase the time to first exacerbation, reduce antibiotic use and improve the quality of life. Long-term clinical benefits have been shown in patients with bronchiectasis and CF. Currently, inhaled mannitol has regulatory approval to be an add-on treatment in patients with CF. In addition to the clinical benefits, the fast delivery of mannitol to the airways and the fact that mannitol can be transported anywhere makes it an agreeable treatment.

http://tinyurl.com/zuxgq5c

Information For People Who Travel On Airlines

In December of 2011, the Transportation Safety Administration (TSA) introduced a toll-free helpline that answers questions for fliers with disabilities and medical conditions. Disabled travelers may call ahead about screening policies, procedures and what to expect at the security checkpoint. The purpose of the new helpline is to inform passengers with disabilities about certain policies before they fly so they may properly prepare for travel. The helpline may be helpful for people with cystic fibrosis who must travel with durable medical equipment, portable breathing machines, needles, liquid solutions (inhalant medications, insulin etc.) and other medical equipment.

Travelers are encouraged to call at least 72 hours prior to a flight. People with CF and other medical conditions may call the "TSA Cares" toll-free number at: 1-855-787-2227. The helpline is available Monday through Friday, 9am-9pm (Eastern Time) and is closed on all Federal holidays.

Individuals also may find information on traveling with special medical needs on the TSA website at:
https://www.tsa.gov/travel/special-procedures

CFRD

The 1-hour oral glucose tolerance test glucose and insulin values are associated with markers of clinical deterioration in cystic fibrosis. Coriati A, Ziai S, Lavoie A, Berthiaume Y, Rabasa-Lhoret R. Acta Diabetol. 2016 Jun; 53(3):359-66.

Cystic fibrosis (CF) is associated with the emergence of CF-related diabetes (CFRD). CFRD is associated with increased risk of accelerated weight and/or lung function loss (clinical degradation). A negative association was observed between the 60-min oral glucose tolerance test (OGTT) glucose value and pulmonary function (FEV_1), whereas 60-min OGTT insulin values were positively associated with BMI. Patients with high G60 values displayed lower FEV_1 than patients with low G60 values. Thus, both insulin and glucose values at 60-min OGTT are associated with indicators of clinical degradation in adult patients with CF. Future prospective analyses are essential in establishing the clinical utility of these indicators.
http://tinyurl.com/hpr2dvw

Glucose Fluctuations Are Not Modulated by the Proportion of Calories from Macronutrients or Spontaneous Total Energy Expenditure in Adults with Cystic Fibrosis. Ziai S, Coriati A, St-Pierre D, Chabot K, Desjardins K, Leroux C, Richter MV3, Rabasa-Lhoret R. Can J Diabetes. 2016 Jul 7. [Epub ahead of print]

CF-related diabetes (CFRD) is the most common complication of CF, and its presence increases morbidity and mortality in patients. Patients with CF (with and without CFRD) have potentially harmful glucose fluctuations. Carbohydrate intake and exercise have been shown to affect glycemia. The authors hypothesized that the proportion of energy from carbohydrates and total energy expenditure (TEE) would influence glucose fluctuations in adults with CF. They concluded that TEE and the proportion of energy from carbohydrates did not affect glucose fluctuations in adults with CF.

http://tinyurl.com/hrlonwr

FYI

Pain and its clinical associations in individuals with cystic fibrosis: A systematic review. Lee AL, Rawlings S, Bennett KA, Armstrong D. Chron Respir Dis. 2016 Feb 12

Pain is recognized as a clinical complication in cystic fibrosis (CF), but the prevalence, characteristics and clinical associations of this co-morbidity have not been systematically reviewed. The pooled prevalence of pain in adults with CF was 77 percent and in children was 42 percent. Common regions of pain included back, abdomen, chest and limbs. In children and adults, pain was associated with a poorer quality of life (QOL) and significant interference with treatments. Pain is a common problem in both children and adults with CF. It has negative clinical associations with QOL and the ability to successfully undertake treatment.
http://tinyurl.com/hm55a5j

Short-Term Effect of Different Physical Exercises and Physiotherapy Combinations on Sputum Expectoration, Oxygen Saturation and Lung Function in Young Patients with Cystic Fibrosis. Kriemler S, Radtke T, Christen G, Kerstan-Huber M, Hebestreit H. Lung. 2016 May 4

Exercise and chest physiotherapy are integral components of cystic fibrosis (CF) care. Exercise followed by physiotherapy has an additive effect on sputum production in participants with CF and leads to improved oxygen saturation. Exercises with increased ventila-

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YOU CANNOT FAIL

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

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

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

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tion combined with mechanical vibration seem to be most efficient.
http://tinyurl.com/jmzgp5a

Supplementation of ursodeoxycholic acid improves fat digestion and absorption in cystic fibrosis patients with mild liver involvement. Drzymała-Czyż S, Jończyk-Potoczna K, Lisowska A, Stajgis M, Walkowiak J. Eur J Gastroenterol Hepatol. 2016 Jun 28(6):645-9

Ursodeoxycholic acid (UDCA) supplementation is recommended for cystic fibrosis (CF) patients with associated liver disease. However, its effect on fat digestion and absorption is not known. The researchers found that UDCA supplementation seems to enhance lipid digestion and absorption in pancreatic-insufficient CF patients with mild liver involvement. This finding points toward the potential impact of UDCA supplementation on nutritional status in CF patients with liver disease and underscores the often overlooked role of factors other than pancreatic enzymes on digestion and absorption of fats in CF.
http://tinyurl.com/z7zg25h

Novel Computed Tomography Scoring System for Sinus Disease in Adults with Cystic Fibrosis. Sheikh SI, Handly B, Ryan-Wenger NA, Hayes D Jr, Kirkby SE, McCoy KS, Lind M. Ann Otol Rhinol Laryngol. 2016 Jun 29. [Epub ahead of print]

There is no easy-to-use scoring system for computed tomography (CT) scans of the sinuses that is specific to cystic fibrosis (CF). The authors proposed a simple and easily implemented scoring system to quantify severity of sinus disease in adults with CF. This scoring system for CF sinus disease was validated. Parameters included maxillary opacification, nasal obstruction, lateral nasal wall displacement, uncinate process absence/demineralization and presence/absence of mucocele. Thus, a novel and easy-to-use CT scoring system for CF sinus disease in adults was validated with inter- and intrarater reliability. This new CF sinus disease-specific scoring system can be used by clinicians, surgeons and radiologists.
http://tinyurl.com/hbjd57b

Colonoscopic screening shows increased early incidence and progression of adenomas in cystic fibrosis. David E. Niccum, Joanne L. Billings, Jordan M. Dunitz, Alexander Khoruts. Journal of Cystic Fibrosis. July 2016Volume 15, Issue 4, Pages 548–553

Colorectal cancer is an emerging problem in cystic fibrosis (CF). The goal of this study was to evaluate adenoma detection by systematic colonoscopic screening and surveillance. The authors found that early screening and more frequent surveillance should be considered in patients with CF, due to early incidence and progression of adenomas in this patient population.
http://tinyurl.com/hl5pl2o

Airway inflammation in mild cystic fibrosis. Eckrich J, Zissler UM, Serve F, Leutz P, Smaczny C, Schmitt-Grohé S, Fussbroich D, Schubert R, Zielen S, Eickmeier O. J Cyst Fibros. 2016 Jun 10. [Epub ahead of print]

Airway infection and inflammation play major roles in the progression of cystic fibrosis (CF) lung disease. In patients with mild disease, airway inflammation is a clinically relevant and often underdiagnosed feature. Lung function, sputum cell counts, and cytokine profiles in CF with mild disease might be different in patients with and

Genetic Mutation Information Resource

Collaborators from several institutions around the world and the U.S. Cystic Fibrosis Foundation are excited to announce that a new resource – CFTR2 – is now available for public use! This is the result of an international research collaboration to provide information about specific cystic fibrosis gene mutations to patients, their families, researchers, health professionals and members of the general public. We hope that you will find the information useful.

The website is available at www.cftr2.org. Once you have reviewed the website, please take a few minutes to complete the user-satisfaction survey located in the blue box "How can you help us improve the website?" in the "Quick Links" section of the left margin. Your responses will help us improve the website. Please feel free to contact cftr2@jhmi.edu with any comments, questions or suggestions, but please note that we are unable to answer any questions about the medical care of individual patients, since we are the research team that helped develop the website and not a clinical care team.

without involvement of small airway disease (SAD). This study demonstrated that patients with CF with mild disease defined by lung function might be further endotyped according to their involvement of SAD. In patients with CF and SAD, airway neutrophilic inflammation is more pronounced and is in part distinct from that seen in patients without SAD.
http://tinyurl.com/jqu8b3a

The changing prevalence of pulmonary infection in adults with cystic fibrosis: A longitudinal analysis. Kay A. Ramsay, Harpreet Sandhu, James B. Geake, Emma Ballard, Peter O'Rourke, Claire E. Wainwright, David W. Reid, Timothy J. Kidd, Scott C. Bell. Journal of Cystic Fibrosis. Article in press.

Increased patient longevity and aggressive antibiotic treatment are thought to impact on the microbial composition of the airways of adults with cystic fibrosis (CF). In this study, the authors sought to determine if a temporal change in the airway microbiology of adults with CF has occurred over time. Longitudinal analysis of sputum microbiology results was undertaken on patients attending a large adult CF center. Clinical status and health outcomes of transitioning patients were also assessed. Results showed a decrease in the prevalence of Pseudomonas aeruginosa, Staphylococcus aureus, Burkholderia cepacia complex and Aspergillus spp. Improvements in lung function among transitioning patients infected with P. aeruginosa were observed. Thus, it may be concluded that over time, a decline in the prevalence of many CF airway pathogens occurs. Significantly, an incremental improvement in lung function was reported for transitioning patients with current P. aeruginosa infections.
http://tinyurl.com/jvbqnuv

Reduced survival in adult cystic fibrosis despite attenuated lung function decline. Claire Keating, Armeen D. Poor, Xinhua Liu, Codruta Chiuzan, Daniel Backenroth, Yuan Zhang, Emily DiMango. Journal of Cystic Fibrosis. Article in press.

There is limited data on disease progression and survival in adult diagnosis cystic fibrosis (CF). This study evaluates change of lung function over time and rates of death/lung transplant in adult diagnosis CF. Annual rate of change of FEV_1 predicted over five years differed by diagnosis age group. Pseudomonas aeruginosa infection was associated with faster rates of lung function decline in all age groups. Lung function appears to decline at a slower rate in adult diagnosis CF. However, particularly in those with low lung function at diagnosis, rates of death or transplant in adult diagnosis CF after 10 and 15 years is not negligible.
http://tinyurl.com/j63blfx

BACTERIA

Longitudinal study of Stenotrophomonas maltophilia antibody levels and outcomes in cystic fibrosis patients. Wettlaufer J, Klingel M, Yau Y, Stanojevic S, Tullis E, Ratjen F, Waters V. J Cyst Fibros. 2016 Jun 23. Epub ahead of print

Previous studies have shown an association between higher Stenotrophomonas maltophilia antibody levels and decreased lung function in patients with cystic fibrosis (CF). The purpose of this study was to assess the serologic response to S. maltophilia over time and to determine whether changes in antibody levels could predict clinical outcomes. It was determined that S. maltophilia antibody levels may be helpful to identify individuals at risk of

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CLUB CF ONLINE

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

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

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

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

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exacerbation who may benefit from earlier antimicrobial treatment.
http://tinyurl.com/zkhyhmk

Clinical outcomes in cystic fibrosis patients with Trichosporon respiratory infection. Esther CR Jr, Plongla R, Kerr A, Lin FC, Gilligan P. J Cyst Fibros. 2016 Feb 27. Epub ahead of print]

Relationships between clinical outcomes and novel respiratory pathogens such as Trichosporon are not well understood. Therefore, respiratory cultures from CF patients were screened for novel pathogens Trichosporon and Chryseobacterium as well as other pathogens over 28 months. Relationships between microbiologic and clinical data were assessed. The presence of Trichosporon, but not Chryseobacterium, is associated with greater declines in lung function. Treatment of Trichosporon-infected patients was associated with improved lung function.
http://tinyurl.com/j8vcq74

PSYCHOSOCIAL

Disease disclosure in individuals with cystic fibrosis: Association with psychosocial and health outcomes. Adrienne P. Borschuk, Robin S. Everhart, Michelle N. Eakin, Devin Rand-Giovannetti, Belinda Borrelli, Kristin A. Riekert. Journal of Cystic Fibrosis. September 2016Volume 15, Issue 5, Pages 696–702

This study aimed to quantify cystic fibrosis (CF) disclosure and examine associations between disclosure and psychosocial and health outcomes. Participants completed measures assessing disease disclosure and psychosocial outcomes. Data from chart reviews and pharmacy records were obtained. Participants were more likely to disclose to romantic partners and close friends than to casual friends, bosses or co-workers. Participants reported more comfort discussing CF with and doing treatments in front of romantic partners and close friends than other groups. Disclosure was associated with higher social support, social functioning and medication adherence self-efficacy. Lower lung-function was associated with disclosure to bosses and co-workers.
http://tinyurl.com/gn5anjb

Psychological resilience and intolerance of uncertainty in coping with cystic fibrosis. Horst Mitmansgruber, Ulrike Smrekar, Bianca Rabanser, Thomas Beck, Johannes Eder. Journal of Cystic Fibrosis. September 2016 Volume 15, Issue 5, Pages 689–695

Anxiety and depression are lower than to be expected in a considerable portion of cystic fibrosis (CF) patients. This outcome might be a result of substantial resilience and/or tolerance of uncertainty in coping with adversity. Research into resilience in cystic fibrosis is in its infancy. 57 adult CF patients participated in the study during their routine medical checkup. In addition to regular psychological assessment, the Intolerance of Uncertainty Scale (IUS) and the Resilience Scale (RS) were administered. Remarkably, resilience (personal competence and acceptance) was clearly elevated, whereas intolerance of uncertainty was comparable to healthy reference groups. Thus, CF patients in this study seem to be particularly resilient rather than cognitively avoidant. At this stage of research, fostering personal competence in CF patients is most promising in improving quality of life.
http://tinyurl.com/zof2bar ▲

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.

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American Organ Transplant Association (AOTA): Phone: 1-713-344-2402 http://aotaonline.org/default.aspx
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