

Look For The Silver Lining

By Kathy Russell

In my opinion, maintaining good mental health is essential to maintaining good physical health. I believe that once one gets into what I call a “funk,” it is so easy to get into a downward spiral that ultimately leads to physical health problems. If one feels tired, cold, blue, sad, uninterested or whatever you may call it, it is difficult to keep up with all of the things that are necessary to keep us well.

I am fortunate in that I was born happy and I have stayed that way most of my life. Rarely have I gotten truly depressed. Being ill repeatedly and having to miss a lot of school didn't depress me. Finding out that I had CF didn't depress me. Even overhearing the doctors telling my parents that I wouldn't live another six months didn't depress me. (I just figured that those doctors didn't know what they were talking about and that I'd show them by outliving all of them...and I have.) Getting ill and having to miss

one whole section of my nursing school didn't depress me. (After all, I was able to make up that unit at the end of my normal rotations.) Having



KATHY RUSSELL

my father die the week that Paul and I got married didn't depress me. It made me terribly sad, but I had other things to do and couldn't let my emotions take over.

The first time that I truly was depressed was when I had to quit working for a living. I had been getting ill more and more frequently. I was missing work and was on the verge of being fired for non-attendance. (This was long before there were laws to prevent such treatment.)

Suddenly I felt as if I had no value. It seems as if everyone measures others by what they do and how much they earn. I was too young to be retired and yet I was unable to work. To add to my feelings of inadequacy, my claim for Social Security Disability was denied. Not only was I not working, I wasn't even contributing any income to our household. I really felt that I was a loser.

I spent a few months trying to get rid of my negative feelings. At that

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EDITOR'S NOTES

Winter seems to be hanging on with a vengeance. By now it seems to me that spring should be here. Soon, I hope. At least we have daffodils and camellias blooming. The flowering fruit trees are starting to bloom, too. C'mon spring!

We have some new directors to introduce. **Amy Sylvis** and **Rachel Steinman** have joined the board. You may read about them on page 14. Welcome to both of you.

Also in this issue, we introduce a new column which replaces "Parenting." It is called "Family Matters" and is about all aspects of families. Two women are authoring it. They are **Molly Pam** and **Cindy Baldwin**. Molly introduces Cindy to us in this issue. Next issue, Molly will be the author.

The Focus topic of this issue is Maintaining Mental Health With CF. **David Tarnow** writes of his perspective of CF and mental health. **John Mercer** continues the discussion in his column, "Financial Solutions," as he describes the importance of being financially healthy to maintain mental health. I add in a peek at my take on depression and happiness on the front page.

In "Ask The Attorney," **Beth Sufian** answers readers' questions about colleges and the Americans With Disabilities Act, receipt of Social Security overpayment notices, and asset limits for receiving SSI.

Isabel Stenzel Byrnes writes in "Spirit Medicine" of healthy boundaries and the importance of maintaining them for our well-being with CF. We find **Jonathan Miller** being interviewed in "In The Spotlight." He is an advocate for CF and shares his love of many things. In the "Poetry Corner" **Linda Stratton** writes of darkness and light in relation to CF.

Once again, **Laura Tillman** has gathered many items about a variety of topics in "Information From The Internet."

You can find announcements about the 2018 CFRI Educational Conference and 2018 Adult Retreat on pages 30-31. Be sure to check out the information about the Lauren Melissa Kelly Scholarship applications on page 9.

Take a look at our upcoming Focus topics on the facing page. Perhaps one of them will pique your interest and you will write for us.

We have some sad news to relay. As we were ready to go to press, our treasurer, **Stephanie Rath**, died. We send our love and understanding to her family and friends. We will have more information about Stephanie in our next issue.

Stay healthy and happy,

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - AbbVie, Gilead Sciences, Kroger Specialty Pharmacy and Two Hawks Foundation in Memory of Dr. Lisa Marino; Pearl Sustaining Partners - Boomer Esiason Foundation, Cystic Fibrosis Foundation; Diamond Sustaining Partners - Marina Day, Trustee of the McComb Foundation, Nancy Wech (in memory of daughter, Lauren Melissa Kelly & in honor of son, Scott Kelly).

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

London Biotech Uses Unique Double Mechanism of Action to Treat Cystic Fibrosis

Verona states their Phase 2a data provide grounds to further develop their drug, RPL554, for the treatment of cystic fibrosis. The drug uses a unique mechanism of action that allows it to target the disease in multiple ways. RPL554 is an inhaled treatment that acts as both a bronchodilator and an anti-inflammatory agent, a feat no other available treatment has been able to match. The drug achieves these two functions through a unique mechanism of action. By using a dual inhibitor, the drug can target two disease-related enzymes (PDE3 and



LAURA TILLMAN

PDE4) simultaneously.

Other treatments for cystic fibrosis are in development as well. Galapagos and AbbVie are collaborating on small

molecule drugs for cystic fibrosis that could restore the function of the CFTR chloride channel, which is defective in cystic fibrosis. French biotech Antabio was awarded \$8.9M (€7.6M) by CARB-X to develop small molecule antibiotics against *Pseudomonas aeruginosa* infections, a common risk to cystic fibrosis patients. Unlike Verona's, these treatments have a single function or target. Researchers from the University of Zurich have found a new chloride channel that could be used as an alternative drug target to CFTR in developing new treatments.

<http://tinyurl.com/y87mz9sr>

CF Foundation Supports Phase 3 Trial of AeroVanc, Inhaled Antibiotic for MRSA Infections, with \$5M Award

The Cystic Fibrosis Foundation Therapeutics (CFFT) has given an award worth up to \$5 million to Savara Pharmaceuticals to support the continued development of AeroVanc, an inhaled antibiotic intended to treat

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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

Spring (current) 2018: Maintaining Mental Health With CF.

Summer (August) 2018: What Is Most Important To You When A Disaster Strikes? (Submissions due June 15, 2018.) What are the things that you know you must have with you in the event of a disaster, either natural or man-made? Where do you keep these essential items? What plans have you made for surviving when basic services are interrupted?

Autumn (November) 2018: Changes We Experience As We Age. (Submissions due September 15, 2018.) What changes have you noticed in your body and health as a result of aging? How do you approach your aging so as to handle it with ease?

Winter (February) 2019: Organ Transplants. (Submissions due December 15, 2018.)



ASK THE ATTORNEY

Questions From Our Readers, Answers From Our Attorney

Questions related to college, Social Security overpayments and bank accounts are answered in this issue's column. The questions were posed by many *CF Roundtable* readers in the past three months. No reference is to a specific reader's situation. Nothing in this column is meant to be legal advice about a specific situation but is meant to be only information.

The CF Legal Information Hotline (CFLIH) can answer questions related to Social Security benefits, Medicare, Medicaid, health insurance, employment and education. If you have questions please e-mail: CFLegal@sufian-passamano.com to schedule a time to speak with the CFLIH.

1. Question: Is a college professor allowed to opt out of the Americans with Disabilities Act (ADA) or the Rehabilitation Act of 1973, Section 504, in relation to students who have a disability such as CF?

Answer: NO!

Adults with CF attending college or graduate school are reporting an increase in discriminatory actions taken by colleges and graduate schools, especially when the student who has CF misses class due to illness.

The ADA and the Rehabilitation Act provide that modifications to policies and procedures must be made for a person with a disability including providing reasonable accommodations to the student. A student who has CF and misses class due to illness can ask for an accommodation to complete work missed after he or she returns to school. If a student is unable to return to work but has spent some time in the classroom, the student could ask for an incomplete and the chance to complete work when he or she is able to do so.

In addition, the student should not suffer negative consequences because of absences due to illness. For example, a professor cannot say he will reduce the student's grade by 50 percent because the student missed class due to illness. The student must request the reasonable accommodation/modification of policy at the time the accommodation/modification is needed. There is no requirement that the student disclose a disability to a professor before there is a need for a reasonable accommodation or a modification of a school policy. Once the reasonable accommodation request is made by the student, the school can require documentation from the treating physician regarding the need for the absences.

Some colleges tell students that a professor can decide to "opt out" of the law. This is not true. A person cannot opt out of following the man-

dates of the ADA or the Rehabilitation Act. A professor cannot decide to ignore the mandates of federal law. These are NOT optional laws.

Many college students with CF go to their college's Office of Students with Disabilities before or when they arrive at the college or graduate school. The Office of Students with Disabilities (Office) staff should discuss accommodations the student needs or may need in the future. The Office staff should help the student obtain the needed accommodations and provide information on what to do if the need for a reasonable accommodation arises in the future. Unfortunately, many students with CF are reporting that their Office of Students with Disabilities is not helpful if they become ill and cannot attend class.

At least 10 *CF Roundtable* readers who have asked questions in the past three months report that the Office of Students with Disabilities tells the student with CF that each professor can decide if the student will be accommodated. The ADA and the Rehabilitation Act do not allow a professor, teacher or any individual to "opt out" of following the mandates of federal law. Individual professors cannot decide to ignore their legal duties to accommodate students with disabilities.

If a student with CF is told a professor is "opting out" of federal law and the Office of Students with Disabilities will not help, the student can go to speak to a dean or the president of the university.

Students should be aware of their rights under the law and should advocate for themselves when a college fails to provide reasonable accommodations needed by the student with CF.



BETH SUFIAN

2. Question: I work at a CF Center and many of our patients are receiving Social Security overpayment notices. Is there a reason for the increase in overpayment notices? What can people do to make sure their Social Security benefits do not stop due to an overpayment of benefits in the past?

Answer: Social Security has been stepping up its efforts to find and collect overpayments from those who receive either Social Security Disability benefits or SSI benefits. Social Security can find that a person was overpaid and request repayment of the overpayment at any time. This means a person who had an overpayment five years ago can be required to pay the overpayment back to Social Security, even if Social Security has recently discovered the overpayment.

An overpayment of benefits occurs when the person who receives Social Security benefits is not eligible for benefits. One way a person can have an overpayment is if the person worked and made an amount of money from work activity that is over the allowable amount a person can work part time and still receive benefits. Another way a person can have an overpayment is if the person who receives SSI benefits has over the allowable amount of assets. SSI requires a person have less than \$2,000 in assets if the person is an individual and less than \$3,000 if the person is in a family consisting of the person with CF and one or more people.

The Social Security Disability benefit program does not consider assets. It is important to know which benefit you are receiving so that you do not go over the asset limit if you are receiving SSI benefits.

For example, if a person who is not married receives SSI benefits, the person cannot have more than \$2,000

in assets. A person who receives SSI benefits can own one car and one house and not have the car or house count toward the asset limit.

If the person has \$2,200 in a bank account, then the person has gone over the asset limit and should not receive SSI benefits. The problem is that often Social Security will not discover the person has gone over the asset limit until 12, 24 or even 32

months later. Social Security can determine that the person was not eligible for ANY SSI payments after the date he or she went over the allowable asset amount.

If 12 months have gone by and the person has been receiving \$750 a month in SSI, then the person could have an overpayment of \$9,000. A person can appeal the overpayment determination if he or she thinks Social Security has made a mistake and he or she did not go over the asset limit. There is a limited time to file an appeal. Another option is to file a Waiver Request and request Social Security waive the overpayment. The person must complete paperwork and explain why he or she thinks Social Security should waive the overpayment.

If a person owes the overpayment, then the person can ask Social Security to set up a payment plan so the person can pay Social Security back over a period of time.

If a person does not agree to a payment plan or pay the overpayment in full, Social Security can stop all Social Security payments until the

withheld benefits pay Social Security back for the overpayment amount. Social Security can take a tax refund to pay back some or all of an overpayment. Social Security can also garnish wages of the person who owes money until the overpayment is paid.

3. Question: I am on a bank account with my mom. Does Social Security count half of the money as mine to determine if I meet the SSI asset criteria?

Answer: If a person's name is on a bank account with someone else, the total amount in the bank account counts toward the SSI individual asset limit of \$2,000.

To reduce money in the account to under the \$2,000 limit, the money has to be spent down on things for the person who receives SSI. The person needs to keep receipts for the items purchased. If the person uses the money to pay bills, the person should retain a copy of the payment stub from the bill. Making copies of checks is also helpful. Social Security can ask for receipts at a later date. A Social Security representative is able to see on a computer if money has been taken out of a bank account.

When a person takes the money out of his or her account and gives the money to someone or puts the money under a pillow, Social Security can find that a fraudulent transfer of funds has occurred. If the person was receiving SSI benefits, the SSI benefit could stop. If the person is applying for SSI, Social Security can impose a five-year period during which the person cannot receive SSI benefits. ▲

Beth is 52 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.

“Some colleges tell students that a professor can decide to “opt out” of the law. This is not true.”



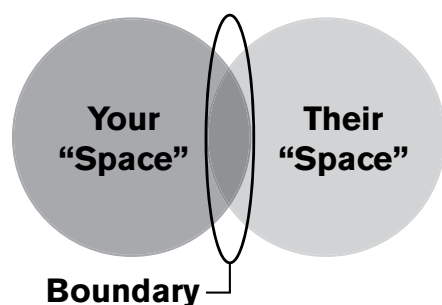
SPIRIT MEDICINE

Healthy Boundaries For The Spirit

By Isabel Stenzel Byrnes

In this “Spirit Medicine,” I’d like to ponder a topic central to the health of our Spirit. This topic pertains to what the Spirit needs, wants and knows is right for itself. This topic is relevant to all relationships and all aspects of mental health. This topic is about healthy boundaries.

In relationships, there is always a Self and there is Other; there is You and there is Me. The line between two people is what I call a boundary. Like this graphic, it is the intersection where two people relate and connect to each other:



We all have comfort zones with our boundaries. Maybe you’ve met people who like to come really close and talk inches from your face. I know that makes me uncomfortable because their germs are too close and they are invading my space. We also have people who subtly and not-so-subtly want something from us that we don’t want to, or can’t, provide. Our boundaries are always being tested. A major source of stress is when boundaries are not respected. This stress hurts the Spirit and causes distress for our mental health. Speaking up, setting limits and saying no all are important ways to maintain our boundaries.

Our boundaries are often informed by our ethnic, societal and family cul-

tures. In some families, family boundaries are loose; each person does his or her own thing and there is little communication and overlap. This is called disengaged. In other cases, families are “enmeshed.” We’ve all seen families where each member tells each other what to do and are arguing all the time; everyone gets caught up in the drama.

As an identical twin with CF, my sister, Ana, and I grew up with very few boundaries between us. We regularly told each other what to do and had no filter in sharing direct opinions with each other, about each other. Though we loved each other dearly, this lack of boundaries led to routine fighting, as we thought we could control each other as if one twin belonged to the other. This dynamic is common in twins, but as we grew older we learned that maintaining harmony meant distancing from each other a bit and respecting each other as two separate people. Life was more

pleasant that way.

Sometimes, the happiness of one person depends on the happiness of those in the family. I’ve seen a CF parent and CF child become so bonded that the parent feels almost like they have CF. That is called love, and this codependency works to aid in the survival of the child. But then comes guilt and unreasonable responsibility for the CF child’s well-being. As the child grows, a lack of boundaries can prevent the CF child from developing his or her own identity. In other words, having CF and dealing with the fear of losing someone to CF can really challenge our boundaries within families. One of the most profound things I’ve heard from a mom of a CF child was, “I *finally* realized it was her disease, not mine.”

In the CF culture, our boundaries are often tested on a regular basis. We have to develop our comfort level with how open we want to be with others about our illness, how much we want to let others in to know our business or help us out. As adults, sometimes we want privacy in how we are dealing with CF (even from our parents or spouses), and sometimes we want to share what’s going on. We have to work with healthcare providers to determine how much to tell them or how friendly we become with them. Loose boundaries can lead to certain expectations and disappointments if those expectations are not met.

CF can sometimes make us feel insecure. Some of us fear that people won’t like us because we are sick. We might be susceptible to giving in to what others want, just so we are included like everyone else. I’ve known friends with CF who got caught up in



ISABEL STENZEL BYRNES

the drinking scene because all their friends were doing it. People with CF cannot be manipulated into doing what others want of them; we must be self-directed to take care of our bodies.

One of the most important aspects of healthy boundaries is having strong self-confidence. A confident person can say no and not care what others think. A confident person can let people down and know it's right for her or him. A confident person can withstand complicated emotions that arise when boundaries are challenged.

According to the book, *How to Be an Adult* by David Richo, healthy boundaries require us to trust our intuition, to heed our own choices, to respect and love ourselves, to honor our own integrity and to have a personal standard that we commit to. Maintaining boundaries is one of the hardest things we can do. Someone else's charm, seduction and flattery can manipulate us. Altering our plans and choices to give in to other people's wishes is generous and selfless, but it can also lead to resentment, anger, powerlessness and even victimization.

Richo states that your boundaries are intact in a relationship when you:

1. Have clear preferences and act on them.
2. Recognize when you are happy/unhappy.
3. Acknowledge moods and circumstances around you while remaining centered (live actively).
4. Do more when that gets results.
5. Trust your own intuition while being open to others' opinions.
6. Live optimistically while co-working on change.
7. Are satisfied only if you are thriving.
8. Are encouraged by sincere ongoing change for the better.
9. Have excited interest in self-enhancing hobbies and projects.
10. Have a personal standard that,

albeit flexible, applies to everyone and ask for accountability.

11. Appreciate feedback and can distinguish it from manipulation.

12. Relate only to partners with whom mutual love is possible.

Just for the comparison, Richo also states you *give up* your boundaries in a relationship when you:

1. Are unclear about your preferences.
2. Do not notice unhappiness since enduring is your concern.
3. Alter your behavior, plans or opinions to fit the current moods or circumstances of another (live reactively).
4. Do more and more for less and less.
5. Take as truth the most recent opinion you have heard.
6. Live hopefully while wishing and waiting.
7. Are satisfied if you are coping and surviving.
8. Let the other's minimal improvement maintain your stalemate.
9. Have few hobbies because you have no attention span for self-directed activity.
10. Make exceptions/alibis for this person for things you would not tolerate elsewhere.
11. Are manipulated by flattery so that you lose objectivity.
12. Keep trying to create intimacy with a narcissist.

All people with CF deserve to be healthy and we deserve to be mentally healthy. I do believe boundaries develop as we mature. They are a result of getting to know oneself and learning what works for you. CF complicates the intersection between you and others. I encourage you to do an inventory of your own relationships and use Richo's list to determine whether these boundaries are working for your Spirit. ▲

Isabel is 45 and has CF. She lives in Redwood City, CA, with her husband, Andrew.

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.





FAMILY MATTERS

NEW
COLUMN

Happy Mother's Day To The Ones Who Struggle

By Cindy Baldwin, with an introduction by Molly Pam

Welcome to the new “Family Matters” column, formerly called “Parenting.” This new column will contain two perspectives on parenting, family and my new favorite term “family building.” The first is from Cindy, a mother to a spirited five-year-old. The other is from me (Molly), a young woman in the process of deciding whether and how to have children. To me, family building implies so much more than just the decision to have kids. It can encompass the whether and how, but it also includes building a family with nieces and nephews, friends, and parents and siblings. No two perspectives or families are alike, and we hope to bring you our thoughts on what family means to us as we alternate columns in the coming issues. We are excited to join the staff at *CF Roundtable* and share our stories. This issue, Cindy will share her thoughts on Mother's Day, a day that can be both so joyous and painful for women with CF. ~Molly

These days, Mother's Day is one of my favorite days of the year. My husband is great at gifts and surprises and my five-year-old loves to get adorably into the action. But it wasn't always that way.

Growing up, I would tell everyone that I wanted ten kids, including a set of twins—just like the grandmother I was named for. I loved babies, little kids, older kids. I could think of nothing more wonderful than spending my

life raising a large, loud, happy family.

I spent most of my childhood ignorant of the more serious ramifications of life with cystic fibrosis. It wasn't until I was a young teen and (ironically enough) my family was spotlighted in *Redbook* magazine that I first read the phrase “average life expectancy”—and along with the understanding that CF wasn't a static disease, that I wouldn't be able to maintain my good health for

my whole lifetime, came the realization that motherhood would be a struggle for me. I was likely to face fertility problems in my future, and pregnancy would put enormous stress on my body. Even if I overcame those challenges, I grew to understand, I would probably not have the physical resources to parent more than a few children at most. I was devastated. I had always been energetic and ambitious, and the

list of things I wanted to do with my life was long—but “mother” had always been first and foremost on that list. More than anything else on earth—more than wanting to write, more than wanting to have a career in music, more than anything I'd ever wanted—I knew that I was meant to be a mother.

“Mothering has always been the core of who I am, and learning as a teenager that there would be so many roadblocks in my path to motherhood shattered me.”

Mothering has always been the core of who I am, and learning as a teenager that there would be so many roadblocks in my path to motherhood shattered me.

I spent a long, long time in those early teenage years seeking comfort about my yearning for motherhood. I tried to make my peace with the idea that I might never be a mother, or that I might never bear children from my own body, or that if I was ultimately blessed with children I might have only one or two—nothing like the full house I had always dreamed of. I struggled with those feelings off and on for a decade—through high school, college, and early marriage. I got married young and would have loved kids right away, but for the first few years my health was unstable enough my husband and I didn't feel comfortable trying to get pregnant. Once my health finally lev-



CINDY BALDWIN

eled out and we felt good to go ahead, I experienced a further year and a half of infertility—something that was not terribly surprising, as I'd always had very infrequent cycles and when my estrogen and progesterone were tested, they came back at levels so low it was almost menopausal.

We considered adoption, but the agency we were interested in had a firm rule that you couldn't be trying to conceive while using their services, and we weren't ready to give up on pregnancy as an option. Finally, four years after we had gotten married, I got pregnant unexpectedly right after beginning Kalydeco. (I was one of about three women who got pregnant almost immediately after Kalydeco hit the market in 2012; at the time, the only pregnant women further along than me were a month or two ahead at best, so it was pretty uncharted territory!)

This March, my daughter turned five, and Mother's Day has been a joy since she was born. But every May, I find myself thinking constantly of the women (and men!) who aren't feeling joyful on Mother's Day—the ones who are in the place I used to be, feeling those same difficult, painful feelings as they see others celebrate the happiness of mothering, a happiness they may

never get to experience. I think, too, of the women who are mothers, but still struggle with the holiday—seeing it as a reminder of how imperfect their mothering is, something that, in my opinion, is a particular struggle for us CF mothers, who are often unable to do so many “normal” mom things with our kids. And every Mother's Day, I find myself wanting to sit down with these women who struggle, look them in the eye, and say:

I see you.

I see you watching the family chaos all around and aching for a child of your own.

I see the grief in your Facebook posts and comments about Mother's Day—or how you take yourself off social media that day, go radio silent so you don't have to witness the pain of all that joy.

I see the way your heart aches for the baby, or child, or teenager, or adult, who isn't with you anymore.

I see the way you so often feel inadequate, like your disease makes you incapable of being the kind of mother you'd like to be. (I know I do!)

I see how hard you work to be gracious, to smile, to keep the bitterness out of your heart.

I see how you sometimes cry alone

in your house when you get that baby shower invitation.

I see you because I have, so often, been you. Not all of you—there are many struggles associated with motherhood that I've never experienced. But the ones that I have experienced? They've sometimes felt like they might rip my heart in two.

Nothing about parenting with cystic fibrosis is easy, and often, celebrations of motherhood can bring that difficulty to the surface. So if you are one who's struggling this Mother's Day, please know that so many of us who are in different places on the journey are thinking of you, remembering vividly the pain we've felt in our pursuit of CF motherhood.

We see you. ▲

Cindy is 29 and has CF. She lives just outside Portland, Oregon, with her husband and spitfire of a daughter (who never makes CF care easy!). Her debut novel, Where the Watermelons Grow, will be published on July 3, 2018, by HarperCollins Children's Books. You may contact her at: cbaldwin@usacfa.org

Molly is 30 and has CF. She lives in New York City. You may contact her at: mpam@usacfa.org

Applications For The Lauren Melissa Kelly Academic Scholarship Due June 30th And October 15th.

CF Roundtable offers the LMK Scholarship award each semester. Nancy Wech, Lauren's mother, established this scholarship in Lauren's name and memory. The academic scholarships of up to \$2,500 are awarded to adults with cystic fibrosis who are pursuing career certifications, associate, bachelor or graduate

degrees. Please visit our website for the LMK Scholarship criteria and application form:

<http://www.cfroundtable.com/community-outreach.united-states-adult-cystic-fibrosis-association-scholarship/>

Please direct any questions to: scholarships@usacfa.org.

IN MEMORY OF
LAUREN
MELISSA KELLY



FOCUS TOPIC

MAINTAINING MENTAL HEALTH WITH CF

Don't Focus On The Numbers: Mental Health And CF, An Altered Perspective

By David Tarnow

As I've gotten older, the effects on my mental health from having CF have become more pervasive, and it has now become something I've had to learn to manage with a similar sort of dedication and attentiveness as I do my physical symptoms. I was very fortunate for most of my young life to live what I used to call a "normal life." When I was diagnosed at the age of three, my parents decided that they were not going to raise me in a bubble. I believe that played an integral role in my psychosocial development and allowed me to have a childhood free from self-labeling and seeing myself as different from my peers. What made this mindset easier at the time was that my physical health was more or less unaffected by the disease. Aside from taking enzymes and doing treatments, my quality of life was not all that different than that of my peers.

As a freshman in high school, I acquired my first road bicycle. With the encouragement of my aunt (an avid cyclist herself), I decided to make the sport a commitment by creating a fundraiser for the CF Foundation. It was known at the time as "Dave's Cycle for a Cure." I was fortunate to have a wonderful support group in my family, friends and the local CF chapter. The framework for the fundraiser was that of a letter writing campaign wherein I shared the practical realities of living with CF, the need for continued donations to help find a cure and the prospect of being able to "save lives one mile at a time." Each year, from 2004 to 2008, my family and I organized a multi-day cycling tour each summer in various

locations across the country. In return for all of the gracious donations that we received, we would blog our often whimsical journeys for the entertainment of donors and to document our efforts. This was also in the days before blogging was "hip," as it is today. The overall effect of conducting this fundraiser over several years was raising hundreds of thousands of dollars for the CF Foundation and receiving awards and accolades for having done so. Although this work was incredibly gratifying, I am not sure that I truly grasped the scope of it all throughout. I was proud to have accomplished such a feat, but I also felt that my struggle with the disease was not nearly as difficult as others'.

I would frequently receive comments and letters from donors and others commending me on my efforts and telling me how inspirational I was.



DAVID TARNOW

I felt undeserving of said praise because, frankly, it was relatively easy. Then, between my senior year of high school and my freshman year in college, my lung function dropped from 108 percent to 67 percent within a period of about three months. What happened? This drastic decline shocked me. Up until that moment, I had not had to overcome significant adversity with regard to my CF, and I was in no way prepared, despite all of my purported strength, to handle the emotional and mental effects of such traumatic news. In very short order, my thoughts became dominated by fears, uncertainty and doubt. I was finally confronted with my own mortality.

Throughout college, most, if not all, of my decisions were subconsciously influenced by my fears of mortality. Instead of approaching life from a perspective of abundance and love, as I had done in prior years, my approach to life and living became one of coming from a place of lack and of fear. Rather than living life intentionally, I went into auto-pilot and was just going through the motions – just doing what I was supposed to do – school, work, treatments, medications etc. I was in denial, and my coping mechanism was to seek perfection in all aspects of my life. This pursuit, however, led me to a perpetual cycle of dissatisfaction.

After graduating college and being in the professional world for a couple of years, at a routine clinic visit, I cultured a bug that I had not ever cultured before. I was told that the treatment for this infection would be long (potentially years, including IVs), difficult and possibly unsuccessful. Unfortunately, my care team at the time was not equipped

“ With the help of medication and therapy, I slowly began the process of acceptance. ”

with the knowledge or skill set to treat me, and there was no standardized protocol for doing so at the time. I began a prescribed therapy and got worse and worse. Within a couple months of starting therapy, my lung function proceeded to drop from 67 percent to the low 30s. It was at that moment that after years of denial and putting on an emotional facade of being “okay,” I lost it. I broke down in front of my doctor, feeling more hopeless than I had ever felt in my life. After resisting taking mental health medication for my entire life, in that moment, I surrendered.

With the help of medication and therapy, I slowly began the process of acceptance. I made great efforts to internalize the mantra, “Life is not a sprint; it is a marathon.” In other words, it did not matter how fast I got to the finish line. What was more

important was that I was able to endure. I started to accept the fact that, ultimately, I was not in control and that my disease had become a part of my identity, and that was okay.

I made efforts to rekindle a sense of joy, optimism and strength in my life, but there were still unresolved emotions. Particularly, I harbored feelings of being less than worthy of being loved. My peers were fostering very successful careers, getting married and starting families. For me, however, being compliant with my treatments, getting rest and avoiding acute illness were my top priorities. The difference in the way our lives looked was a stumbling block for me. As Teddy Roosevelt once said, “Comparison is the thief of joy,” and I found this to ring true most of the time. I had to learn to be gentle with myself. When in community, I allowed myself

to be vulnerable with everyone about what my life looked like with CF. I made them aware of why I made the decisions I made and the struggle of self-care (mental and physical). In so doing, I began to find that “Those [people in my life] who matter don’t mind, and those who mind don’t matter.”

One piece of advice that has always stuck with me came from one of my past CF doctors who told me, “Don’t focus on the numbers [referring to my FEV₁]. The numbers are only a small part of the whole story. What is important, however, is whether or not you are able to do the things you love with the ones you love.” What a wise old sage. I have spent too many years in my short life focusing on the numbers, letting them consume and define me and my ability. I hope that you do not do the same. Life is for living. You are worthy; you are stronger than you think you are, and you are loved. ▲

David is 29 and has CF. He lives in Erie, CO. You may contact him at dtarno1@gmail.com

Correction

In the Winter 2018, issue of *CF Roundtable*, the printers inadvertently placed a photo of **Julie Desch** in the spot for the photo of **Julie Browning Ice**. We regret the error and are printing the correct photo and caption here, with our apologies.



PHOTO BY RONNA

JULIE BROWNING ICE



In Memory

Sandi Alvaro Dick, 30
Fairmont, WV
Died November 17, 2017

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

Send to:

CF Roundtable
PO Box 1618,
Gresham, OR 97030-0519

E-mail to:

cfroundtable@usacfa.org

time, we lived above the ocean in Rancho Palos Verdes, California. Each day I would go across the street to the cliffs above the ocean. A couple of feet down from the edge, there was a little ledge where I could sit and just watch the ocean. I listened to the sounds of the world and watched the sea birds, seals and other sea animals and just let my mind unwind. Every day that I sat like that I felt better than the day before. The sun felt good and helped me to get better.

Eventually, I got to the point that I felt okay with going to one of the pools at our condo and visiting with others who gathered there. Many of us liked books and we would spend time discussing current books we all were reading or some topic that had caught the interest of one or more of us.

I was the youngest of the people who joined these discussions. Some of them had worked and some had not. Some still were working but came around on their time off. It really didn't matter to anyone that I was no longer

working. To them, I just was a neighbor who was sharing the pool and the conversation. My attitude about myself really improved from all that time in the sun, by the water and visiting with nice people.

I still have periods of a kind of depression that is none too deep. I get

library books in the return slot. Paul gets out of the car and does it, since it is difficult for me to get my concentrator out of and back into the car. I detest being unable to just open the door of our home and run out to get some flowers from the yard. I resent having to watch out for my oxygen hose all the time.

On the other hand, I am delighted that I have oxygen available and that I have portable oxygen, too. I have much more freedom than I might have and I am alive. It helps to remember that my concen-

trator gives me much needed oxygen and that it allows me to be a viable part of life.

For the first 50 years of my life, I had serious sinus disease that caused me terrible pain. At times it felt as if my teeth needed to be pulled to relieve the pressure in my head. My eyes felt as if they were being pushed right out of their sockets. It was depressing to hurt all of the time. That pain was relieved by sinus surgeries.

My favorite way of avoiding depression is to always try to find the bright side of any situation.

depressed about how so many people in the world have not learned to get along with others. I know that this particular type of depression isn't the result of CF, but it does have an effect on my mental health and hence on my physical health. I do what I can by sharing love and caring where I am able.

I get depressed over being tethered to an oxygen hose all of the time. I really dislike being unable to just hop out of the car and run up to drop my

persistent methicillin-resistant *Staphylococcus aureus* (MRSA) lung infection in individuals with cystic fibrosis (CF). Specifically, the award will support testing of the clinical effectiveness of AeroVanc (vancomycin hydrochloride inhalation powder) in a pivotal Phase 3 (NCT03181932) trial called AVAIL. AeroVanc is the first inhaled antibiotic aiming to treat MRSA lung infections in CF patients. In a Phase 2 study in CF patients with persistent MRSA infection (NCT01746095), AeroVanc was found to reduce MRSA density in sputum and showed encouraging trends of improvement in lung function. AVAIL's primary outcome will be the mean absolute change from base-

line, or study start, in the volume of air a person can forcibly exhale after a deep breath in one second (forced expiratory volume percent predicted or FEV₁) at week four and week 20 of treatment. <http://tinyurl.com/y6uhkg19>

Proteostasis Therapeutics Announces Positive Clinical Results from Studies of PTI-428, PTI-801 and PTI-808 in Healthy Volunteers and Patients with Cystic Fibrosis

Proteostasis Therapeutics, Inc., announced positive study results across all three of the company's CF pipeline programs. These include a study in CF subjects for PTI-428, a cystic fibrosis transmembrane conductance regulator

(CFTR) amplifier; interim data for a study in CF subjects for PTI-801, a new generation CFTR corrector; and studies in healthy volunteers for PTI-808, a CFTR potentiator, and the combination of PTI-428, PTI-801 and PTI-808. The results support the goal of studying the Company's novel CFTR modulators in doublet and triplet combinations in CF subjects. <http://tinyurl.com/y9u4wdp6>

Proteostasis's Proprietary Triple Combination Therapy Study Protocol Receives Endorsement and High Strategic Fit Score from the Largest Cystic Fibrosis Patient Advocacy Groups in the U.S. and Europe

After my last sinus surgery, which was 23 years ago, I lost my senses of smell and taste. At first I found that to be depressing. I had always loved cooking. A large part of cooking is smelling and tasting what is being made. Suddenly, I found myself unable to taste anything. I had to guess on seasonings and flavorings, where I wasn't using an exact recipe. I got my husband to help me with adding the correct amount of whatever spice or flavoring I was using. We managed. I did really miss tasting food. I still yearn for the taste of a real root beer float. If I were to drink one today, mostly what I would taste is sweet and I don't particularly like sweet. Oh, well. At least I am able to eat and can taste something now and then.

We used to plant a huge garden. We grew corn, green beans, lettuces, radishes, spinach, herbs, squashes, cucumbers, tomatoes and various other vegetables. I enjoyed weeding, watering, harvesting and preserving the bounty of our gardens. I would get quite tired after spending a day harvesting and canning or freezing. It was well worth it, when I could look at my

pantry shelves and see the jars of beautiful produce. I loved being able to control how much salt or sugar went into anything I preserved. My jars of peaches and pears were delicious with a light syrup that was my own recipe. It allowed one to really get the flavor of the fruit. My tomatoes had no salt on them, only water. They were delicious. Being able to remember all of that keeps me from being depressed that I no longer have the energy it takes to do all of that work.

Getting a diagnosis of breast cancer (I know, it isn't CF-related) didn't cause me to be depressed, either. I figured that "this is just another thing to deal with." I had bilateral mastectomies four-and-a-half years ago. Clothes don't fit as well as they used to, but I am alive and that is more important.

I have a twisted spine that causes me almost continuous pain. Because of that, I find that I sit a lot more than I did when I was younger. I am not sure that this is CF-related. It may be just because I am old. I would prefer to be more active, but I run out of energy too quickly to do much that takes real work.

I keep my legs elevated to keep my ankles from swelling. (I don't want to have "cankles," as unkind folk call them.) Since I sit with my feet up, I find that working on my computer or doing jigsaw puzzles affords me activities that are pleasant and doable. Finding puzzle pieces that were being elusive makes me happy. Editing the material for this newsletter really gives me pleasure. I truly enjoy doing all that is required in editing.

My favorite way of avoiding depression is to always try to find the bright side of any situation. Even if it is a life-altering event, there still may be an upside to it. Focusing on only the negative aspects of any situation is almost guaranteed to cause depression. Finding the "silver lining" behind any "black cloud" can help to keep from letting depression win. Look on the bright side and stay much happier is my recommendation.

Until next time, stay healthy and happy. ▲

Kathy is 73 and has CF. She is Managing Editor of CF Roundtable. Her contact information is on page 2.

Proteostasis Therapeutics, Inc., announced that its triple combination clinical study protocol has received endorsement and a high strategic fit score from the Therapeutics Development Network (TDN) and the Clinical Trial Network (CTN). The TDN and CTN are the drug development arms of the Cystic Fibrosis Foundation (CFF) and the European CF Society (ECFS), respectively. The study is designed to investigate the safety, tolerability and pharmacokinetics (PK) of the co-administration of PTI's cystic fibrosis transmembrane conductance regulator (CFTR) modulators, PTI-428, PTI-801 and PTI-808, and will also assess lung function (FEV₁).

<http://tinyurl.com/yafyzk7g>

Cystic fibrosis: Discovery of a key molecule for improving treatments

Researchers at the University of Montreal Hospital Research Centre (CRCHUM) have identified a promising solution to improving treatments offered to patients with cystic fibrosis. Adding molecules called quorum-sensing inhibitors to current drugs not only reduces bacterial production of certain harmful residues but also restores the efficacy of existing treatments on the cells of cystic fibrosis patients. Over the past decade, two prescription medications have been offered to eligible patients: Kalydeco for those with rare

mutations (fewer than four percent of patients), and Orkambi for the more frequent mutation (79 percent of patients). Unfortunately, these two drugs have only limited efficacy. Researchers are trying to understand why. Current drugs act to correct the defect in the CFTR protein. In a sterile in vitro environment, Kalydeco and Orkambi treatments work well. But in real life, the lungs of sick patients are colonized by bacteria, *Pseudomonas aeruginosa* in particular. Much evidence has indicated that the bacteria could interfere with the treatment. Due to this study, the substances released by the bacteria that could reduce the effi-

Continued on page 15

Meet A New Director

Hello! My name is Rachel Steinman, I'm 29 years old, and I'm super excited to be a member of the USACFA board.

I was diagnosed with CF at the age of 16 and have been very fortunate to have had fairly stable health throughout my life thus far.

No matter one's level of health, USACFA is an important outlet for our community. CF is a complicated disease and it affects every patient in a unique way. For me, having an online community has helped me both learn more about my disease and feel less isolated in the process.

I graduated from the University of Miami with degrees in Journalism and

Sociology in 2009 and I spent a year volunteering with AmeriCorps shortly after. A few years into my career, I

decided to quit my job and move to Tel Aviv, Israel, for a change of lifestyle and a marketing position with a tech company. I moved back home to New York after a year to be close to my family.

I grew up on Long Island and currently reside in NYC with my boyfriend where I continue to work in digital marketing. I enjoy cooking, traveling with my boyfriend and spending time with friends and family.

I believe I've been able to maintain good health with the help of amazing doctors, a positive attitude, lots of acupuncture and cupping therapy and a very loving, supportive family.

I can be contacted at: rsteinman@usacfa.org. ▲



RACHEL STEINMAN

Meet A New Director

Hello! My name is Amy Sylvis and I am so grateful to be a member of the USACFA board. I have been an avid reader of *CF Roundtable* since the late '90s and I am forever grateful for how much I have learned over the past two decades from the publication.

I fiercely believe that all people with cystic fibrosis should have access to the best care and latest knowledge – and I'm thrilled to be able to contribute to this prestigious organization. My specific passions include hemoptysis, aspergillus, CFRD and decreasing quality of treatment variation across CF centers in the United States.



AMY SYLVIS

I am 36 years old. I was diagnosed at six months old. I earned my Bachelor of Science in Business and Bachelor of Arts in International Relations from the University of Southern California as well as my Masters in Business Administration from USC. I have worked full time, mostly in biotech and pharma, which continues to be my passion despite my health forcing me to leave work in 2017.

In my spare time you can find me reading nonfiction, watching college football and traveling. My husband and I were married in August 2017, and we live in Los Angeles, CA, with our little cocker spaniel/dachshund mix. ▲

cacy of the treatments has been determined. The researchers tested, in vitro, the effect of adding a quorum-sensing inhibitor from the furanone family. This molecule, by reducing the bacterial production of harmful residues, was able to maintain treatment efficacy in the cells of patients with cystic fibrosis.
<http://tinyurl.com/ybdcvj8>

Cancer gene plays key role in cystic fibrosis lung infections

PTEN is best known as a tumor suppressor, a type of protein that protects cells from growing uncontrollably and becoming cancerous. But according to a new study PTEN, working with another protein, CFTR, also keeps lung tissue free and clear of potentially dangerous infections.

The findings, published in *Immunity*, explain why people with cystic fibrosis are particularly prone to respiratory infections—and suggest a new approach to treatment. Recent findings suggested that cells with CFTR mutations have a weaker response to bacteria, reducing their ability to clear infections and augmenting inflammation. The research team discovered that when PTEN is located on the surface of lung and immune cells, it helps clear *Pseudomonas* bacteria and keeps the inflammatory response in check. But PTEN can do this only when it's attached to CFTR. And in most cases of cystic fibrosis, little CFTR finds its way to the cell surface. As a result, the duo fails to connect, and *Pseudomonas* runs wild. However, the latest generation of cystic fibrosis drugs push mutated CFTR to the cell surface, with the aim of improving chloride channel function and reducing a buildup of mucus. The new findings suggest that it might be beneficial to coax nonfunctional CFTR to the surface as well, since even abnormal CFTR can work with PTEN to fight infections. The study also raises the possibility that PTEN might have something to do with the increased risk of gastrointestinal cancer in cystic

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SUSTAINING PARTNERS



Supported by a grant from AbbVie

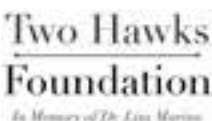
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Thanks To The Boomer Esiason Foundation

CF Roundtable's New Pearl Sustaining Partner

We would like to thank The Boomer Esiason Foundation for its continued support now at the Pearl Sustaining Partner level. A special thank you goes out to BEF volunteer Jerry Cahill for helping make this grant possible. Because of this support, we can provide all of our CF Roundtable programs such as:

- CF Roundtable Newsletter
- CF Roundtable Blog: <https://goo.gl/iqsNhc>
- The Speakers Bureau: <https://goo.gl/WNdtDX>
- Awards to individuals who make a difference in the CF community: <https://goo.gl/6LfGXG>
- Support to college students tackling CF and an academic schedule: <https://goo.gl/AvksXN>

The Boomer Esiason Foundation helps support the CF community via its programs including:

- Scholarships – numerous scholarship opportunities available
- Lung Transplant Grant Program – a program that covers transportation, housing and other expenses related to transplant not covered by insurance.
- You Cannot Fail – a motivation program that empowers people with CF

- Hospital Bags – goodie bags provided to CF patients of all ages during hospital stays
- CF Podcasts – podcasts covering a wide variety of CF-related topics, produced by Jerry Cahill
- CF Wind Sprints – short videos by BEF volunteer Jerry Cahill with tips for living with CF
- Club CF – an online forum where people with CF can share their stories
- Gunnar's Blog – a personal blog of Gunnar Esiason, Boomer's son, who has CF
- Team Boomer – a system that encourages people with CF to be active by participating in events and helping to fund-raise
- Bike 2 Breathe – an annual 500-mile bike tour to raise awareness for the importance of exercise with CF
- CF Century Rides – a personal goal of Jerry Cahill's. Jerry is determined to do a century ride (100-mile bike ride) in all 50 states for CF!
- CF: Live By Example – a pilot program where people with CF who are living, breathing and succeeding will ensure parents of newly diagnosed children that CF is only a bump in the road, not a death sentence.

For more information on The Boomer Esiason Foundation please visit: <https://www.esiason.org/>



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United Network For Organ Sharing 1-800-355-7427

THROUGH THE LOOKING GLASS



PHOTO BY RAY LOCKETT

Augie's Clinic

She doesn't ask
About the productivity
of my cough
or what meds I'm taking.
My weight is meaningless to her.

But she willingly keeps me company
on those sleepless nights
full of coughing fits.
And extra snacks are just a bonus
to her foraging nature.

She takes no interest
in the results of blood work
or sputum cultures.
Readings from the peak flow
and glucose meters tell her nothing.

For, she already knows
the best way to counteract
unpleasant procedures
a game of tug-o-war,
a belly rub or squeaky toy
makes everything seem a little better.

She has her own methods
for counteracting bad days,
those of shortness of breath,
fatigue and no appetite.

And sometimes all I need
is what she has to offer
the silliness of a jester,
an extra nap on the couch,
soft fur soothing
skin that has felt
too much harshness for one life.

-C. Martinet, 2003

FROM OUR FAMILY PHOTO ALBUM...



LINDA STRATTON (MIDDLE) WITH NEW NEIGHBORS AND FRIENDS, LOUELLA (LEFT) AND ANNETTE (RIGHT), AT THEIR NEW SENIOR COMMUNITY IN LOUISVILLE, COLORADO.



MOLLY PAM AND HER HUSBAND, ADAM COHEN, HANGING OUT IN NYC.



AMY (SYLVIS) AND JOEL (GUTIERREZ) ON THEIR WEDDING DAY IN AUGUST 2017, MALIBU, CALIFORNIA.

PHOTO BY ANNA DELORES PHOTOGRAPHY



**MAHON, KATE AND CINDY
BALDWIN, JULY 2017.**

**JONATHAN MILLER PRE-
SENTING AT A HEALTH-
CARE PANEL ALONGSIDE
CONGRESSWOMAN
ELIZABETH ESTY, CT,
DISTRICT 5.**



**RACHEL STEINMAN AND
HER BOYFRIEND, RAIDEL
LAZCANO, IN NYC THIS
PAST DECEMBER.**





ACTIVE FOR LIFE

Community Interview With Justin Seleska

By Aimee Lecointre

Hey friends! I hope this issue finds you all doing well. I don't know about you, but I am so looking forward to warmer weather ahead. As I'm writing this (end of March) it's snowing outside! Spring, where you at? Personally, I am much more motivated to exercise and be active during the warmer months, probably because sunshine is my favorite vitamin and I love being active and working out outside.

This issue, I want to introduce you all to my friend Justin Seleska. Justin and I share a love for all things San Diego, staying active to help manage our health with CF and remaining optimistic. However, we don't share a love of running. He's a runner, I'm definitely not. He once ran a half-marathon a month for a year, impressive! Even though I don't personally enjoy running, I'm super motivated by anyone who does what they can to move.

1) Hey Justin, thanks for being my guest this issue. Can you tell us a little about yourself?

My name is Justin Seleska. I'm 38 years old with CF. I grew up in Redlands, CA. In my early 20s I set out for a better life healthwise and overall by moving to San Diego and have remained here ever since. I am surrounded by an amazing support system of family and friends that keeps me in balance. This includes my sister who has started running half-marathons with me as well.

2) I don't blame you for not leaving San Diego, I miss it daily! So tell us, what role does exercise and staying

active play in your life with CF?

It allows me to clear my lungs out of "junk" and hopefully that means fewer infections and fewer antibiotics (oral and IV). I have always been pretty active growing up, too. I played baseball when I was younger and moved to wrestling while I was in high school.

3) I know you're a runner and one year you ran one half-marathon a month, so rad by the way. How did you get started with running?

I started with running while I was laid off from work one time in late

2012. I was looking for something to fill my time, so I decided to take up running. I registered for my first half-marathon which would occur in March 2013 and I haven't looked back since.

4) What keeps you motivated to keep running/exercising?

My health and wanting to keep my lungs in as good a shape as I can. I am going to do my best with what I can control to keep pushing these breathers.

5) What are your tips for anyone with CF who wants to get started in running?

For anyone who starts running, I would first recommend researching and getting good shoes. Nothing will kill running faster than poor shoes. If you really want to go for distance, I would recommend finding a running

program that you realistically think you can do. But other than that, just get outside and get yourself moving, whether it's just down the street, around the block or out for distance.

6) I love that advice and often give it myself, just move! I know you also like to hike and have participated in the Xtreme Hike for CF fundraisers for the CFF. Aside from hiking and running, do you have any other ways you like to stay active?

This year will be my third Xtreme Hike. I try to go out and surf and sometimes go to the gym. I also just like to take a nice relaxing ride on the boardwalk on my beach cruiser. Plus I also live in an area of San Diego where a lot of things are walkable, so I try to walk most places when I can.

7) San Diego makes it easy to stay active for sure. Let's chat now about one of my other favorite topics, food!

"I was looking for something to fill my time, so I decided to take up running."



JUSTIN SELESKA (RIGHT).

What are your favorite pre- and post-run meals?

Before runs, usually a banana and some kind of bar. After runs, I usually down a protein shake. After a major run, though, I usually go out with the family to an eating establishment and enjoy whatever protein I can find and also a celebration beer.

8) Any words of encouragement or anything else you'd like to share with us?

I would encourage every CFer to "just be you." I know that's cliché and may or may not work for you. At the same time, try something that does work for you. I would just recommend doing what you can to keep your lungs as healthy as you can. Like I mentioned before, just walk up the street, around your house etc. Just to keep moving

and keep the lungs going.

9) One last thing, I know you just started a podcast. I'm pumped about this because you and I have pretty similar mindsets. Want to tell us a little bit about it?

I started a podcast to get a voice out there of someone who is living life to the fullest with CF. I want to tell it how it is and that all is not lost in the battle. Yes, I understand that unfortunately a lot of people still lose that battle early, but with recent advances many more people will be able to live a "normal" adult life without CF controlling every aspect.

Thank you so much Justin for chatting with me and sharing a bit of your story with our amazing community!

Now CF Roundtable readers, I need your help. I love interviewing different

members of our community about the ways they stay active and I hope you enjoy it too. Here's where I would love your help. Is there something different you'd like to see in this column about exercise/movement and CF? Something you'd like to see more of? Please feel free to e-mail me and let me know, I'd love to hear from you! You can e-mail me at aimee.lecointre@gmail.com ▲

Justin is 38 and has CF. Currently, he lives in San Diego, CA. He works at an electronics company. In his free time he enjoys running, going to concerts, spending time at the beach, pretending he can actually surf and hiking.

Aimee is 32 and has CF. She lives in Salt Lake City, UT, with her husband, two pugs and a cat. You may contact her at aimee.lecointre@gmail.com.

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fibrosis patients.
<http://tinyurl.com/yahb75kh>

Defect in zinc supply mechanism affects pathology of intractable pulmonary diseases

Researchers have made it clear that abnormalities of the ion channels ENaC and CFTR reduce the body's ability to deliver zinc ions (Zn^{2+}) to lung epithelial cells. (One of the zinc ion transporting factors, ZIP2, is an important mechanism for mRNA splicing to function correctly.) In addition, the researchers also revealed that CF lung epithelial cells have abnormalities in mRNA ligation, which, as a result, causes overproduction of mucus genes. In other words, the disease developed due to an overproduction of mucus which was caused by an insufficient zinc supply to lung epithelial cells. The researchers believe that this discovery is the first in the world

that clarifies how zinc is involved in the onset of pulmonary diseases, as well as its effects on the regulation of mRNA, one of the molecules responsible for life as we know it. These results prove that there is abnormality in the zinc transport mechanism itself in obstructive pulmonary diseases. It also suggests that treatment for these diseases cannot be performed merely with zinc supplements. A therapeutic approach that takes the transport mechanisms into consideration also is necessary.

<http://tinyurl.com/y7y8popd>

AND

<http://tinyurl.com/y9atfjw5>

Synspira Announces \$3 Million Development Award from Cystic Fibrosis Foundation

Synspira announced that it has received up to a \$3 million award from the Cystic Fibrosis Foundation to

advance clinical development of its lead candidate, SNSP113, for use in pulmonary complications of cystic fibrosis (CF). SNSP113 is a novel glycopolymer representing a new class of molecules developed to treat infection, inflammation and congestion in the lungs. SNSP113 breaks up bacterial biofilms and weakens bacteria, allowing for the potentiation of antibiotics. In addition, it diminishes mucus viscosity, reducing inflammation and promoting clearance of the lungs.

<http://tinyurl.com/yaryq8b4>

Potential CF Therapy for Bacterial Infections Gets Patent, May Start Clinical Testing

A possible inhalable treatment for antibiotic-resistant bacterial infections in people with cystic fibrosis due to *Pseudomonas aeruginosa* now has a U.S. patent and is being readied for a first

Continued on page 24



IN THE SPOTLIGHT

With Jonathan Miller

By Andrea Eisenman and Jeanie Hanley

In this column, you will get to know a thoughtful and curious person who has a vast variety of interests. He seems to make the most of his time learning about things that pique his interest. While learning about his own health, he tries to be aware of how to treat what might be coming around the bend. It seems to me (Andrea) that he approaches his health in thoughtful ways and shares how you might be able to take a page out of his book. But what impressed me most, initially, was his interest in CF advocacy regarding health insurance—not just for his own gain but to benefit others who have CF and to educate local statewide lawmakers. I hope you all can take away some new ideas to better care for yourselves. Meet Jonathan Miller, our latest star (who also happens to know a lot about astronomy!). Spotlight, please!

What made you such a vocal advocate for healthcare for people with CF?

It was a combination of many things, but mostly because I benefited so much from the Affordable Care Act. My experiences with our healthcare laws were nothing like what I heard from so many different politicians. So I wanted to share my experience in hopes of educating others about how our laws protect me in so many ways, and what would happen to me and others if they were to be changed for the worse.

How did you get started?

My advocacy started with a simple e-mail sent to one of my U.S. Senators. I was then invited to attend a meeting to share my story and, from there, quickly started getting more offers from different politicians and media outlets to do all kinds of speeches,



JONATHAN MILLER AND HIS COUSIN, OLIVIA, PLAYING ROCK-PAPER-SCISSORS.

articles, presentations, even a podcast. I always said yes when asked, no matter how large or small the platform. If I could help shape one person's mind about how they think about healthcare, I wanted to do it. As CF patients, I believe it is our responsibility to let our lawmakers know who we are and what our needs are when it comes to healthcare. Obviously, there is only so much any of us can do at the federal level, but all of us should be in touch with our local representatives at the state level. They influence a lot of healthcare policy, especially when it comes to Medicaid. If they don't know who you are, you aren't being considered when it comes time to vote!

How important is it for people to be involved, if they can, in their healthcare choices?

Because so few people actually understand how health insurance works, it leaves them vulnerable to misinformation. I think everyone

should take the time to learn the basics – premiums, co-pays, deductibles – these are important terms to know that will help anyone better understand their coverage. For patients with CF, unfortunately, I think all of us need to know a lot more than the basics. Because healthcare policies change every year, it's important to know all the treatments that you need and how they get covered. If you don't know these details, you could end up with a plan that does not cover everything you need or is too expensive. The best way to learn is to ask questions and take advantage of the resources that are available to you, especially CF Legal (www.sufianpassamano.com) and Compass (CFF).

Did you see your health improve on disability?

I used to work full time at a university as a program director for fundraising programs. I went on disability because I physically could not work

anymore. I was getting exacerbations much too frequently and symptoms were more aggressive. I was very nervous about going on disability, at first. I was worried about the unknown, and working and earning a living was very important to me. However, being on disability gave me the time I needed to properly take care of myself.

How did you improve your health?

The most important thing I learned, since becoming disabled, is how to distinguish my symptoms and figure out what's causing them. Because my time was no longer consumed with work, I found myself noticing far more of the subtle things my body was doing. I am now much better at sensing the different types of airway obstruction in my lungs and sinuses. For many years I thought that my health was random, that sickness and exacerbations were always arbitrary and just a part of my health. The biggest change in my understanding these last few years has been finding that my condition is shockingly consistent. While I do get sick quite a bit, I've learned that, for me, almost no symptom comes without an identifiable cause. As symptoms begin to show, I can start to troubleshoot and identify the reason. Often it's simply an environmental change – air quality, allergies, contaminated respiratory equipment, changing air purifier filters etc., which requires a simple change. Once I've figured out the likely cause and made appropriate changes, a lot of times I can recover with the right balance of rest, exercise, water, ibuprofen, increased albuterol treatments and airways clearance, and changing my exposure to anything that might be causing my symptoms.

Tracking and treating my symptoms this way takes a lot of work and it's important to not become obsessive. To strike the right balance, I try to be very diligent when symptoms present them-

selves but more hands off when things are normal. Of course, there are limits to how much one can do without medical intervention, so it's always important to stay mindful of those limits and work with your doctors to chart your best path forward. Sometimes I have to be reminded of that, too!

What tricks can you share for others to track their symptoms?

I think it's important for everyone to establish what their health goals are. Once you have those, you can start to figure out what changes you need to make in order to reach them. You can identify patterns, you can start to find causes and continue to make adjustments as you go.

When I am recovering from an exacerbation, I look for small signs of improvement each day. Are my pulse oximetry numbers a little bit better? Is my energy level a little higher? Can I do a little more exercise today than yesterday? As long as these things are improving, then I know I'm on the right track.

What is important to you in life?

One of the unexpected great things about going on disability was that it left me with much more free time. For perhaps the first time in my life, I felt free to answer a simple question – “What do I WANT to do?” Having this freedom revealed a lot about myself and what I value. One of the things I decided to finally spend more time on was astronomy and stargazing. I quickly found myself enjoying learning and found an appetite for other science subjects as well.

A lot of the things I learned about science didn't just change the way I viewed the physical world, but also my understanding of CF. As I learned some basics about subjects like chemistry, and even just air, I started to better understand how my body interacts with the world around me. This

expanded knowledge of air quality and how airborne pathogens spread gave me a lot more insight into preventing illness. Learning a bit more anatomy, especially of the lungs, really helped as well. It gave me new ways to think about how to take care of myself.

What is your philosophy on living?

A life with CF has provided me with the bittersweet awareness of how short life is. It's this awareness that often drives me to do all that I can, for as long as I can, and to try to properly value the opportunities that come my way. I care about living life in a way that encourages new experiences and growth, not just for myself but for others as well. It's the times I'm doing the things I love, with the people I love doing them with, that always bring me the most joy and even moments of freedom from my disease.

What do you enjoy doing?

I play a lot of sports, love playing music and writing songs. I'm a great dancer, when I'm well enough. I didn't understand the true importance of hobbies until I became sicker. I have learned the value of having hobbies I can do at various levels of physical condition. Having things I can do even when I am sick and much less mobile really helps me get through those times.

When I am well, I also enjoy golf, disc golf, bowling, ballroom dancing, billiards and hiking.

From whom do you get support?

I get support from a lot of different people. I rely on each family member and friend in different ways. I have also benefited tremendously from working with a therapist. I recommend that EVERYONE see a therapist at least every once in a while. It's a tremendous opportunity if you are ready to be open and curious. If nothing else, it's nice to

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have someone you can say literally anything to and get out your frustrations without fear of negatively impacting someone you care about. Most of my sessions I just vent! Through the great relationships I have, I don't generally feel lonely, but I do live with a certain sense of isolation. I spend a lot of time with myself and my thoughts, and my experiences are so vastly different from all the other people in my life, I don't have anyone I feel truly understands me. Like my support system, I find that different people understand different parts of me, and so I work hard to keep these people in my life and find time for fun with the people I care about.

How do you handle hardships?

We, as CF patients, get a rare lens through which to witness the world. We experience a spectrum of lows and highs that most others never will. The skills and character of which I am most proud have been cultivated from the hardest, most stressful aspects of my life, and while I certainly wish many of these moments had never come, I'm sometimes surprised, even amazed, with how much I've been able to grow from them. I still work to avoid hardship, but when it must come, I try to welcome it and the lessons it will bring me. ▲

Jonathan is 27 and has CF. He lives in Meriden, CT. You can find him all over New England - in the woods, on the golf course, in bowling alleys, under the night sky

and sometimes on a dance floor. He loves sharing experiences and learning new things. He has drug-resistant Burkholderia cepacia and hopes there will be drugs that can treat these strains of bacteria some day. You can e-mail him at jonathanmiller163@yahoo.com. He will respond to any inquiries... eventually!

Andrea Eisenman is 53 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 54 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.

clinical trial, Novoclem Therapeutics announced. The bacterium *P. aeruginosa* causes chronic lung infections in CF patients. Its antibiotic resistance, coupled with the thick mucus commonly found in CF patients' lungs, makes it difficult to treat *P. aeruginosa* infections. BIOC51 was designed to be inhaled as a dry powder or nebulized solution and penetrate mucus to better reach and eliminate *P. aeruginosa*. It releases nitric oxide in controlled amounts to attack both free-floating bacteria and bacteria growing in biofilms. Having a water soluble, tunable system for releasing nitric oxide created a new paradigm for treating disease and an ability to deliver nitric oxide to a much broader range of locations within the body. Nitric oxide has several properties, including antimicrobial activity. BIOC51 was given qualified infectious disease product (QIDP) status by the FDA in 2017 as a potential treatment for recurrent lung infections caused by bacteria.

<http://tinyurl.com/yclk5d6a>

AIT Therapeutics Announces Further

Data From Its NO-NTM Abscessus Phase 2 Clinical Trial in Nontuberculous Mycobacteria (NTM) Patients

AIT Therapeutics Inc., a clinical-stage biopharmaceutical company focused on developing inhaled Nitric Oxide (NO) for the treatment of patients with serious lung infections and pulmonary hypertension, today announced positive data at Day 81, the end of its NO-NTM Abscessus clinical trial. With just 21 days of treatment, patients still benefitted at Day 81 as shown by 6-minute walk data. Additionally, the anti-microbial effect of NO is apparent with quantitative PCR (qPCR) showing a 65 percent reduction in bacterial load. All patients in the trial were resistant to the standard-of-care for NTM abscessus (MABSC) and had underlying cystic fibrosis. The possibility of using AIT's system to treat patients chronically, at home, is a very promising development.

<http://tinyurl.com/y87ccggg>

Savara Reports Publication of Case Reports of Inhaled Granulocyte-

Macrophage Colony Stimulating Factor for the Treatment of Nontuberculous Mycobacteria

Savara, Inc., reports the publication of two case reports exploring the use of aerosolized granulocyte-macrophage colony stimulating factor (GM-CSF) for the treatment of *Mycobacterium abscessus* (*M. abscessus*), a species of multidrug-resistant nontuberculous mycobacteria (NTM), in individuals living with cystic fibrosis (CF). The case reports show inhaled GM-CSF eradicated or dramatically reduced *M. abscessus* infection, improved clinical outcome and was well tolerated. Molgradex, a proprietary inhaled form of GM-CSF, offers a novel treatment approach for NTM infection by stimulating the human immune system in the lungs with localized delivery of GM-CSF directly into the site of infection. Whereas Molgradex may be effective on its own against NTM infection, it may also be useful in combination with any of the current or future antibiotic regimens. NTM lung infection is a considerable therapeutic challenge due to the

From Darkness Into Light

By Linda Stratton

Cystic fibrosis—my nemesis, threads of bacteria my long-term guests
 Going deeper into infection, breath and light decline into darkness
 Blinds drawn, a sheet over my head, needing to shut out the world
 A desire to be numb, so I sleep
 Overwhelmed by illness, so I sleep
 When sleep eludes, a pill assists, then again I sleep
 PICC line inserted, meds start to flow
 Fingers of life and light filter through the depths of darkness
 Peeking out from under cloak of seclusion, compassion surrounds, cradled in arms of care
 Health and well-being begin to dawn, cognitive of day and night
 Each step a victory as my world opens up, possibilities of normal life
 Thirteen laps equal a mile is my goal, each day getting stronger
 Venturing out, IV pole and all, fresh air and sunlight warm my face
 It's a process, a repeated dance, each time CF controls
 Aware of how precious time in the light truly is
 My heart bursts with thanksgiving and joy
 Thank You Lord

unique ability of these bacteria to evade the normal killing mechanisms of alveolar macrophages, a type of immune cell responsible for killing bacteria in the lungs. GM-CSF is not an antibiotic. Instead, it stimulates the human immune response without targeting the bacteria directly, thus avoiding the problem of inducing antibiotic resistance.
<http://tinyurl.com/y9ece3kf>

Review Board Recommends Continuation of Phase 3 Trial of Anthera's CF-related Digestive Therapy

An independent review board has given a green light to Anthera

Pharmaceuticals continuing a Phase 3 clinical trial evaluating Sollpura's (lipomatase's) ability to treat a cystic fibrosis-related digestive disorder. It was the second time that the Data Monitoring Committee had decided that interim findings were good enough for the RESULT trial (NCT03051490) to continue. The study is comparing Sollpura's and the standard treatment Pancreaze's ability to treat exocrine pancreatic insufficiency. A hallmark of CF is overproduction of thick mucus in several places in the body. One manifestation of this is the pancreas being unable to generate enough of the enzymes the gut needs for

digestion. To overcome this, CF patients can receive pancreatic enzyme replacement therapies, which are commonly derived from pigs. Sollpura consists of non-pig-derived lipase, protease and amylase enzymes. Its synthetic origin reduces the risk of patients developing a pig-related disease or conditions such as gout and kidney impairment. Because Sollpura's formulation is more compact than pig-derived enzyme replacement therapies, it can also reduce the size and number of pills that CF patients must take each day to achieve similar results. Anthera has also developed a powder

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FINANCIAL SOLUTIONS

Mental Health And Financial Health: A Surprisingly Close Relationship

By John Mercer

Mental health is a topic that I take seriously and I think it impacts people more than most understand. I'm one who spends a lot of time in my own head, so I have a great deal of empathy and sympathy for anybody who struggles with mental health. Managing money and being financially astute are not the easiest things to discuss; not always "party conversation." However, it's a conversation that needs to happen and avoiding it may have direct implications on your mental health.

Wrapping one's head around all that comes with being well-educated financially isn't easy, especially if you're having other struggles in life. Interestingly, there is a bi-directional and closely related relationship between finance and mental health. People struggling with mental health have a hard time managing their finances, and people who have trouble with finances often have mental illnesses and bouts of depression. Poor mental health means managing money is more difficult, and worrying about money negatively impacts mental health.

So much of what we do or think about doing, so much of what we're able to do, where we want to travel et cetera is closely related to the amount of money we have. The best piece of advice moving forward is to become as educated as you possibly can about all aspects of your finances. The more informed, knowledgeable and confident you are about finances results in less stress and worry regarding financ-

es. Motivate yourself to become educated sooner rather than later. It will blow your mind how the U.S. economy works and how being prepared financially can change your life.

Debt and mental health

The most significant relationship is between debt and mental health. A meta-analysis from the University of Southampton found that over the

course of 80 large studies, people with debt were three times more likely to experience mental health problems than those who were debt free. Depression, anxiety, anger and countless other disorders were included. A second significant finding: people who committed suicide were eight times more likely to have debt. While causation is unclear, many have speculated that owing money causes high rates of anxiety and stress. Very likely this is because money encompasses so much of our life and is already something we worry about. The issue of wanting to pay something back can triple the stress that managing money already causes.

Because the link is so strong, I want to make everyone aware of the facts

before I talk about solutions. There are some extremely conclusive ways to improve mental health as it relates to debt. First, when you have debt the most important thing to do is to openly acknowledge it and not hide it. Looking for a support system (typically for larger debts) or just letting someone know that you owe them money (typically for smaller debts) can help a great deal. When you owe another person money, just reminding them that you are aware of it can positively impact both parties. Second is to put a structured goal-driven plan together to solve that problem. With a definitive goal-driven plan, the likelihood of somebody having related mental health issues is drastically lower. This may seem counterintuitive, but the more you acknowledge debt and develop a plan, the easier it becomes to manage

“People struggling with mental health have a hard time managing their finances, and people who have trouble with finances often have mental illnesses and bouts of depression.”



JOHN MERCER

both. Recognizing that link between debt and mental health and tackling it with a positive goal-driven mentality empowers you to overcome both.

Planning for the future: ignorance is NOT bliss!

There is a strong degree of variability among people who think they're financially well-educated and people who actually are financially well-educated. Here's a brief example from Forbes. People who "blindly" (not actively looking at fees, investment costs and revenue sharing) invest in a 401k often report being "moderately or well-educated financially." However, the same people who don't actively monitor their accounts often pay 1.8 to 2.1 percent more annually in excessive fees than those actively monitoring. That can account for 10 years of retirement income. For them ignorance is not bliss. Most people are unaware and that's totally okay, it's not their fault; schools don't teach it, families rarely talk about it and the industry is happy with that.

Ignorance is not bliss when it comes to your money. The government and financial industry are predicated on people being unaware of what is happening with their money. You not being well-educated is the "green-light" for banks, financial institutions and government agencies to skim off the top. By the way, all of this is public record, all over the internet and openly discussed within the industry. Don't listen to me. In 1939 Henry Ford said this: "It is perhaps well enough that the people of the Nation do not know or understand our banking and monetary system, for if they did I believe there would be a revolution before tomorrow morning." Henry did well enough by humanity, I think!

A vastly important thing you can do is to look to the future and figure out what your dream life looks like. Then,

you reverse engineer everything before that point to get you where you want to be. You have to try and plan for what you want in your professional life, family, retirement, real estate and a multitude of other areas. Evaluating those is easier when you have a better idea where you want to go. Then, it's about following your plan. You must become so well educated on the paths/options to get you there that you become the expert for yourself. Know where fees, taxes and everything else go in your savings, investments, bank accounts and retirement vessels. If you don't know, or don't ask, many "financial/bank people" will take advantage. Once you know what's happening, there's no going back to where you once were! So plan, plan, plan and learn, learn, learn. Putting an action plan together and setting goals will enable unbelievable things.

What you can do...

I want to briefly talk about solutions to help you combat some stressors that come with managing finances. Each of these could be its own individual article, but I just want to skim the surface:

Plan as far ahead as possible and become educated on everything that it takes to get there. Whether that's real estate, professional goals, retirement/investment portfolios, a piggy bank underneath your bed or family planning. Embrace that challenge of becoming your own expert, it'll pay dividends. If you don't know where you want to be, it's tough to figure out how to get there.

Talk to a fiduciary professional. I emphasize the word *fiduciary*. A fiduciary professional (typically a planner or consultant) is someone who puts your interest ahead of everything else. They are legally and ethically bound by that title and by putting people first. Remember the word *fiduciary*. Often, financial professionals have a skewed view on things and possess ulterior

motives for themselves or their companies rather than having your best interest first. Make sure you find someone you can trust and who will talk openly and honestly. The right person should be able to tell you the pros, cons and costs of everything. Just trusting that "my finance guy is a friend of a friend" is a bad idea. Make sure you can openly discuss things whenever you want. Someone who is open, honest and compares the numbers is usually a good one.

Save, save, save. Invest, invest, invest. If you're reading this, you can become truly wealthy at some point if you start saving early and let your assets compound as they should. Albert Einstein once said: "Compound interest is the eighth wonder of the world. He who understands it, earns it... he who doesn't... pays it." That's not to say you can become wealthy by investing in a piggy bank beneath your bed, but finding an option that saves you the most money and gains good returns can move mountains. Here's the story of Theodore Johnson, a UPS driver who never made more than \$14,000 a year, but he invested just 20 percent of his earnings yearly, started early and invested in the company stock option retirement plan. When he retired in 1952, the compound interest and the money he saved amounted to \$39 million. After taking out money during retirement, it was worth \$70 million when he passed. He credited habitual saving and not wanting to have to work until he died. I don't know about you guys, but I could use \$70 million. If we save early and let compound interest do its work, we could all be hanging out on yachts in 30 years. Fight on. ▲

John is 34 and has CF. He is an investment planner and senior consultant in Pasadena, CA. He is a house party enthusiast and organizational strategist. You may contact him at: jmercerc@usacfa.org.

SIX Ways To PAY IT FORWARD To CF ROUNDTABLE!

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MILESTONES

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

ANNIVERSARIES

Birthday

Molly Pam

New York City, NY

30 on March 15, 2018

Transplant

Zachary Hays, 29

Vernon, CT

Bilateral lungs

1 year on February 3, 2018

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formulation of Sollpura for oral administration that is being evaluated in the Phase 3 SIMPLICITY clinical trial. (NCT02734810).

<http://tinyurl.com/ycv92yyv>

Among Adult CF Patients, Hypertonic Saline Prolongs Mucus Clearance, Study Finds

A single dose of inhaled hypertonic saline (HS) solution increases mucus clearance for at least four hours in adults with cystic fibrosis (CF). A previous clinical trial of 164 patients over the course of one year showed that twice-daily inhalation of 7 percent HS improved lung function, reduced pulmonary exacerbations and decreased the need for antibiotic treatments. So far, none of the HS studies have addressed whether a single HS dose is enough to endure better mucus clearance, or if repeated daily doses are required. Researchers determined mucociliary clearance rates in three separate conditions: no HS, after 15 minutes (acute response) and four hours (prolonged response), and after one dose of inhaled 4 mL of 7 percent HS. Each condition was tested independently and on separate days for each individual. Mucociliary clearance was measured as the mean lung particle clearance following a single administration of aerosolized radiola-

beled particles. Four hours following HS treatment, lung particle clearance increased by 21.8 percent, compared to 13.8 percent among those not given HS. Coughing was similar in the treated and non-treated conditions. Overall results suggest that the clinical benefits seen for HS in CF patients derive from its long-lasting effects rather than repeated short-term effects on mucus transport. The fact that a single dose is sufficient to accelerate mucus clearance in CF patients for at least four hours contrasts with the effect of HS in healthy subjects, where no prolonged improvement was seen.

<http://tinyurl.com/y9oulopz>

Cystic Fibrosis Therapy Acebilustatystic Receives New American Patent

Celtaxsys has obtained a U.S. patent for **acebilustat**, its lead candidate for treating the lung inflammation found in cystic fibrosis. **Acebilustat** is designed to reduce an overabundance of white blood cells known as neutrophils in CF patients' lungs. The high levels are associated with inflammation. By reducing neutrophils, **acebilustat** decreases lung inflammation and dysfunction, without any immunosuppressive effect. Celtaxsys is conducting a Phase 2 clinical trial (NCT02443688) of acebilustat's ability to reduce lung inflammation and preserve lung func-

tion over 48 weeks. The ongoing trial is also assessing the once-a-day oral dose's safety, and patients' ability to tolerate it. <http://tinyurl.com/y6vwtq4n>

CF Studies Report That Ion-Sugar Interaction Can Affect Airway Mucus

The interaction of ions and sugars called glycans can affect the mucus that lines airways. Two studies showed that ion-glycan interaction can affect mucus's structure and fluidity. Glycans are complex sugars that all organisms have. They help maintain cell and tissue structure, play a key role in cell signaling and protect our body from pathogens like bacteria. They are found on the surface of cells and the mucus that coats organs like the lungs, throat and gastrointestinal tract. The body's molecules can have positive or negative charges, and glycans are among the most negatively charged. Although positive and negative charges attract each other, two different ions with the same charge do not interact equally with an ion that has an opposite charge. This difference is called lyotropy. A key implication of these subtle differences is that glycans interact in unique ways with each type of ion and this affects the thickness of the cells' protective layer against pathogens. Lung mucus contains mucins,

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Conference Speakers:

Elaine Chen, M.D.; Rush University Medical Center, Chicago, IL; **CF and Pain**

Reid D'Amico; Ph.D. Candidate at Vanderbilt University, Nashville, TN; **Science, Advocacy and CF Life**

Manu Jain, M.D.; Northwestern University Feinberg School of Medicine, Chicago IL; **The CF Pipeline**

Jasleen Kukreja, M.D. & Rupal Shah, M.D.; UC San Francisco, San Francisco, CA; **Innovations in Lung Transplantation**

Richard Moss, M.D.; Stanford University, CA; **CF Fungi and Pathogens**

Chelsea Toth, D.S.W.; Kutztown University, Kutztown, PA;

Coping with Isolation

Alan Verkman, M.D., Ph.D.; UC San Francisco; San Francisco, CA; **History and Future of CFTR Modulators**

Sabrina Walker; Anchorage, AK; **Running, Adversity and CF**

CFRI-Funded Research Track Speakers:

Guillermo Flores-Delgado, Ph.D.; UC California San Diego; **Secretory and Absorptive Epithelium in Conductive Airways**

Peter Haggie, Ph.D.; UC San Francisco, CA; **Small Molecule Screening to Validate CF-Relevant Drug Targets**

Nam Soo Joo, Ph.D.; Stanford University, CA; **New Insights into Impaired Mucociliary Clearance in CF**

Kenji Okuda, Ph.D.; UNC at Chapel Hill, NC; **Localization**

of Secretory Mucins MUC5AC and MUC5B in Normal Human Airways

AKM Shamsuddin, Ph.D.; UC San Diego, CA; **Pharmacology of CFTR HCO₃⁻ Secretion in Native Small Airways in Health and Disease**

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negatively charged proteins that are sensitive to variations in ions they come in contact with. The understanding of how each type of ion affects the structure, electrical potential and fluid behavior of the airway-surface layers in the lung has direct implications for designing drug targets that influence pulmonary behavior.

<http://tinyurl.com/y8al8g2m>

Garlic: Weapon against lung disease – Study

Scientists have discovered a substance in garlic that could be a weapon against serious complications in cystic fibrosis. The researchers found that the natural garlic compound called ajoene, could prevent bacteria from communicating and collaborating with each other and thus making it easier to kill them. The study showed that ajoene is espe-

cially effective in fighting the bacteria, *Pseudomonas aeruginosa*. Ajoene also prevents the bacteria secreting rhamnolipid, a substance that destroys the body's white blood cells, which are designed to kill bacteria and prevent infections. According to the study, ajoene does not kill bacteria directly, but it inhibits virulence genes controlled by quorum sensing. This disruption prevents the bacteria from coordinating their attacks on the body. In laboratory trials with a biofilm comprised of *P. aeruginosa* bacteria, the research group showed that ajoene in combination with an antibiotic drug kills more than 90 percent of the bacteria.

<http://tinyurl.com/y8k3pah4>

UF Study Links Proton Pump Inhibitor Use with More Frequent Hospitalization in CF Patients

Doctors should frequently re-evaluate the use of protein pump inhibitors (PPIs) for cystic fibrosis (CF) patients as long-term PPI use leads to a higher risk of hospitalization for pulmonary exacerbations. PPI use is believed to cause community-acquired pneumonia (CAP). Even though most CF patients use PPIs to control gastroesophageal reflux (GER), scientists still don't fully understand the link between PPIs and pulmonary exacerbations in CF. In this study researchers investigated that link and the risks it entails. The study is limited, in that it's retrospective and doesn't establish a cause-effect relationship between PPIs and pulmonary exacerbation. Researchers say there's still a possibility that GER itself causes increased pulmonary exacerbation. Yet they point out that the prevalence of

Continued on page 32

GER was similar among hospitalized and non-hospitalized patients, supporting a causative link between PPI and pulmonary exacerbations. Based on their findings, the team suggests that prescribers of PPI therapy should frequently re-evaluate indications and appropriateness of therapy and in the setting of GER consider alternate management modalities such as anti-reflux surgery where appropriate.

<http://tinyurl.com/y7c5ol4e>

Pulmonary Hypertension Does Not Affect CF Patients' Survival, Study Finds

Pulmonary hypertension increases the risk of cystic fibrosis patients' lung function worsening, but is not a predictor of reduced survival. In addition to chronic lung infections and bronchiectasis, CF patients can develop pulmonary hypertension (PH), which can lead to poorer outcomes. In these patients, the main cause of PH is diminished oxygen levels in tissue, which leads to constriction of lung arteries and increased pulmonary blood vessel resistance. The destruction of lung tissue and associated loss of pulmonary blood vessels is another possible cause of PH in CF.

<http://tinyurl.com/yc882dyb>

FYI

Attention deficit hyperactivity disorder symptoms in patients with cystic fibrosis Malena Cohen-Cymberknoh, Tzlil Tanny1, Oded Breuer, Hannah Blau, Huda Mussaffi, Diana Kadosh, Silvia Gartner, Alma Salinas, Lea Bentur, Vered Nir, Michal Gur, Joel Reiter, David Shoseyov, Eitan Kerem, Itai Berger. *Journal of Cystic Fibrosis*. Article in Press.

The occurrence of ADHD symptoms in patients with CF is substantially higher than in the general population and should be recognized as a co-morbidity of CF. As ADHD can impair adherence to therapy, further research is needed to investigate the effect of

ADHD therapy on adherence.

<http://tinyurl.com/ycjbuena>

AND

<http://tinyurl.com/y7hgfbj2>

Is there an association between back pain and stress incontinence in adults with cystic fibrosis? A retrospective cross-sectional study. Jane E. Ashbrook, Carol Shacklady, Sue Johnson, Gillian Yeowell, Peter Charles Goodwin. *Journal Of Cystic Fibrosis*. January 2018 Volume 17, Issue 1, Pages 78-82

Back pain and stress urinary incontinence (SUI) are common in adults with cystic fibrosis (CF). This study aimed to establish whether there is an association between back pain, lung function and stress urinary incontinence and its relative risk. An analysis of data shows an association is indicated between back pain (SF-MPQ and VAS) and SUI in adults with CF. This information is important when developing management strategies in the CF population.

<http://tinyurl.com/y7b95wrs>

Variceal Hemorrhage and Adverse Liver Outcomes in Patients with Cystic Fibrosis Cirrhosis. Ye, Wen; Narkewicz, Michael R.; Leung, Daniel H.; Karnsakul, Wikrom; Murray, Karen F.; Alonso, Estella M.; Magee, John C.; Schwarzenberg, Sarah Jane; Weymann, Alexander; Molleston, Jean P. for the CFLDnet research group. *Journal of Pediatric Gastroenterology and Nutrition*: January 2018, Volume 66, Issue 1, pp 122-127

Cirrhosis occurs in 5 percent to 10 percent of cystic fibrosis (CF) patients, often accompanied by portal hypertension. Three adverse liver outcomes were analyzed: variceal bleeding (VB), liver transplant (LT) and liver-related death (LD) and risk factors for these. VB is an uncommon complication of CF cirrhosis and can be an indication of the diagnosis, but it does not affect all-cause mortality (ACM). Adverse liver out-

comes and ACM are frequent by 10 years after cirrhosis report.

<http://tinyurl.com/y7qwypm6>

The CF-CARES primary palliative care model: A CF-specific structured assessment of symptoms, distress, and coping. Deborah Friedman, Rachel W. Linnemann, Lily L. Altstein, Suhayla Islam, Kieu-Tram Bach, Chelsea Lamb, John Volpe, Caitlin Doolittle, Anita St. John, Patricia J. O'Malley, Gregory S. Sawicki, Anna M. Georgiopoulos, Lael M. Yonker, Samuel M. Moskowitz. *Journal Of Cystic Fibrosis*. January 2018 Volume 17, Issue 1, Pages 71-77

Current palliative care tools do not address distressing chronic symptoms that are most relevant to cystic fibrosis. A CF-specific structured assessment based on a primary palliative care framework was administered. Patients reported numerous physical and psychological symptoms (mean of 10 per patient), with psychological symptoms rated as more distressing. Anxiety (34 percent) and depression (44 percent) were prevalent and correlated with distress attributable to physical symptoms and difficulty with CF self-management, but did not correlate with disease severity. Frequently reported symptoms are not consistently associated with distress, suggesting the importance of individualized evaluation. The CF-CARES (Coping, goal Assessment, and Relief from Evolving CF Symptoms) primary palliative care assessment model provides a framework for patients experiencing chronic symptoms to explore interventional options with their clinicians.

<http://tinyurl.com/y8ww6skp>

PATHOGENS

Emergence and spread of worldwide *Staphylococcus aureus* clones among cystic fibrosis patients. Garbacz, K., Piechowicz, L., Podkowik, M., Mroczkowska, A., Empel, J., Bania, J. *Infection and Drug Resistance*. 22 February 2018 Volume

The aim of this study was to assess the relatedness of molecular types of *Staphylococcus aureus* isolates colonizing cystic fibrosis (CF) patients with their antimicrobial resistance and prevalence of toxin genes. The findings imply that individuals with CF can also be colonized with animal-related ST398 MRSA, and justify constant monitoring of staphylococcal colonization and identification of epidemic *S. aureus* clones in this group.
<http://tinyurl.com/ya2vgh47>

TREATMENTS

Treatment of chronic rhinosinusitis with dornase alfa in patients with cystic fibrosis: a systematic review. Gopi B. Shah M.D., MPH, Linde De Keyser M.D., Joy A. Russell Ph.D., MLS, Ashleigh Halderman, M.D. International Forum of Allergy & Rhinology. 11 January 2018

A major component of sputum in cystic fibrosis (CF) patients is polymerized DNA, a byproduct of degraded neutrophils. Dornase alfa (dornase) selectively cleaves extracellular DNA and reduces the viscosity of sputum. It improves mucociliary clearance and pulmonary function. The benefit of dornase on CF-associated sinusitis is less clear. Therefore, the objective of this study was to systematically review the use of dornase on chronic rhinosinusitis (CRS) in CF patients. Topical intranasal dornase appears to improve sinonasal symptoms in CF patients to a greater degree than saline alone. The impact on other outcomes is less clear. Larger studies are needed to fully elucidate the true efficacy of dornase alfa in the treatment of CRS in CF patients.
<http://tinyurl.com/y8uw3bap>

Effect of oral glycine on the clinical, spirometric and inflammatory status in subjects with cystic fibrosis: a pilot randomized trial. Mario H. Vargas, Rosangela Del-Razo-Rodríguez, Amando

López-García, José Luis Lezana-Fernández, Jaime Chávez, María E. Y. Furuya and Juan Carlos Marín-Santana. BMC Pulmonary MedicineBMC series. 2017;17:206

The clinical, spirometric and inflammatory status of subjects with CF improved after just eight weeks of glycine intake, suggesting that this amino acid might constitute a novel therapeutic tool for these patients. Thus, further studies are warranted.
<http://tinyurl.com/yc6zhrta>

Long-acting inhaled bronchodilators for cystic fibrosis. Cochrane Database Syst Rev

Inhaled bronchodilators are prescribed for 80 percent of people with cystic fibrosis in order to widen the airways and alleviate symptoms. Both short- and long-acting inhaled bronchodilators are used to improve respiratory symptoms. Short-acting inhaled bronchodilators take effect in minutes and typically last for four to eight hours (muscarinic antagonists). Long-acting inhaled bronchodilators also take effect within minutes but typically last for around 12 hours and sometimes longer. The authors concluded that neither long-acting beta-2 agonists nor long-acting muscarinic antagonist bronchodilators demonstrated improvement in the primary outcome of FEV₁. No difference was observed between intervention and placebo in terms of quality of life or adverse events. The quality of evidence for the use of beta-2 agonists was very low. The use of a long-acting inhaled bronchodilator may help to reduce the burden of treatment for people with cystic fibrosis as it is taken less often than a short-acting inhaled bronchodilator, but future trials would benefit from looking at the effects on the primary outcomes (spirometric changes from baseline, quality of life and adverse effects) in the longer term.
<http://tinyurl.com/y7ye8f0z>

The use of an alternate side lying positioning strategy during inhalation therapy does not prolong nebulization time in adults with cystic fibrosis: a randomized crossover trial. Ruth L. Dentice, Mark R. Elkins, Genevieve M. Dwyer and Peter T. P. Bye. BMC Pulmonary MedicineBMC series 2018;18:3. 8 January 2018

Inhalation of nebulized medications is performed in upright sitting to maximize lung volumes. The pattern of deposition is poor for inhaled medications in people with cystic fibrosis. The pattern tends to be non-uniform and typically the upper lobes receive a reduced dose compared to the rest of the lung. One strategy that has been proposed as having the potential to improve homogeneity of deposition is to adopt an alternate side lying position for the inhalation procedure. This study sought to determine whether there is any disadvantage to delivery time of nebulized medications with a strategy of alternate side lying, compared to upright sitting. Results show that side lying during inhalation therapy does not prolong nebulization time. Two-minute periods should provide an equal dose in the two side lying positions.
<http://tinyurl.com/yajqyny7>

Recovery of lung function following a pulmonary exacerbation in patients with cystic fibrosis and the G551D-CFTR mutation treated with ivacaftor. Patrick A. Flume, Claire E. Wainwright, D. Elizabeth Tullis, Sally Rodriguez, Minoo Niknian, Mark Higgins, Jane C. Davies, Jeffrey S. Wagener. Journal Of Cystic Fibrosis. January 2018. Volume 17, Issue 1, Pages 83-88

Pulmonary exacerbations (PEx) are associated with acute loss of lung function that often is not recovered after treatment. The authors investigated lung function recovery following PEx

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for ivacaftor- and placebo-treated subjects. They determined that ivacaftor treatment reduces the frequency of PEx but does not improve on the rate of complete lung function recovery after PEx when compared with placebo. <http://tinyurl.com/ydxglpqs>

Retrospective observational study of French patients with cystic fibrosis and a Gly551Asp-CFTR mutation after one and two years of treatment with ivacaftor in a real-world setting. Dominique Hubert, Clémence Dehillotte, Anne Munck, Valérie David, Jinmi Baek, Laurent Mely, Stéphane Dominique, Sophie Ramel, Isabelle Danner Boucher, Sylvaine Lefeuvre, Quitterie Reynaud, Virginie Colomb-Jung, Prissile Bakouboula, Lydie Lemonnier. *Journal Of Cystic Fibrosis*. January 2018 Volume 17, Issue 1, Pages 89-95

Ivacaftor has been shown to improve lung function and body weight in patients with CF and a gating mutation. Real-world evaluation is warranted to examine its safety and effectiveness over the long term. A retrospective observational multicenter study collected clinical data in the year before and the two years after ivacaftor initiation in patients with CF and a Gly551Asp-CFTR mutation. The clinical benefits of ivacaftor reported in previous clinical trials were confirmed in a real-world set-

ting two years post-initiation, also reducing treatment burden. <http://tinyurl.com/y8l6sns5>

Hypertonic saline has a prolonged effect on mucociliary clearance in adults with cystic fibrosis. Aaron T. Trimble, A. Whitney Brown, Beth L. Laube, Noah Lechtzin, Kirby L. Zeman, Jihong Wu, Agathe Ceppe, David Waltz, William D. Bennett, Scott H. Donaldson. *Journal Of Cystic Fibrosis*. Article in Press

Inhaled hypertonic saline (HS) has been shown to increase mucociliary clearance (MCC) and improve clinical outcomes in adults and adolescents with cystic fibrosis (CF). We previously demonstrated the absence of a sustained effect of HS on MCC in healthy adults and in this study sought to characterize the durability of the MCC response to HS in adults with CF. These results suggest that, in contrast to healthy adults, a single dose of HS has a prolonged effect on MCC in adults with CF, which lasts at least four hours. This may explain its clinical efficacy in this population. <http://tinyurl.com/yc9wkfxv>

Clinical experience with ceftazidime/avibactam for treatment of antibiotic-resistant organisms other than *Klebsiella pneumoniae*. Barbara A. Santevecchi, Tiffeny T. Smith, Shawn H. MacVane.

International Journal Of Antimicrobial Agents. Article in Press

Ceftazidime/avibactam is a newly approved β -lactam/ β -lactamase inhibitor combination with activity against antibiotic-resistant Gram-negative organisms, including many carbapenem-resistant strains. Although this agent may offer a promising treatment option for serious infections with limited alternatives available, clinical experience with ceftazidime/avibactam in treatment of infections caused by multidrug-resistant Gram-negative organisms other than *Klebsiella pneumoniae* is limited. A retrospective case series was performed to evaluate patients treated with ceftazidime/avibactam for infections caused by organisms other than *K. pneumoniae* at our institution over a one-year period. For infections caused by antibiotic-resistant Gram-negative organisms other than *K. pneumoniae*, clinical and microbiological success rates for patients treated with ceftazidime/avibactam were similar to those that have been reported for *K. pneumoniae*. Ceftazidime/avibactam appears to be a promising treatment option for infections caused by a variety of resistant Gram-negative organisms when limited alternatives exist. <http://tinyurl.com/ycz9uwf3>

KB001-A, a novel anti-inflammatory, found to be safe and well-tolerated in

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cystic fibrosis patients infected with *Pseudomonas aeruginosa*. R. Jain, V.V. Beckett, M.W. Konstan, F.J. Accurso, J.L. Burns, N. Mayer-Hamblett, Carlos Milla, D.R. VanDevanter, J.F. Chmiel for the KB001-A Study Group. *Journal of Cystic Fibrosis*. Article in Press

Chronic *Pseudomonas aeruginosa* (Pa) airways infection, exuberant local inflammation and progressive lung function loss are hallmarks of cystic fibrosis (CF). KB001-A is an anti-PcrV

PEGylated monoclonal antibody fragment to the Type III secretion system of Pa. This 16-week study evaluated KB001-A associated effect on time-to-need for antibiotics for worsening respiratory signs and symptoms, as well as safety and treatment-associated changes in symptom scores, inflammatory markers and spirometry. The authors found that KB001-A was safe and well-tolerated and associated with a modest FEV₁ benefit and reduction in select sputum

inflammatory markers (IL-8). KB001-A was not associated with an increased time to need for antibiotics. The lack of efficacy seen with KB001-A may be due, in part, to the low levels of the Type III secretion proteins previously reported in sputum of CF patients chronically infected with Pa.

<http://tinyurl.com/y7gswmth>

SINUS

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Pulmonary aspiration of sinus secretions in patients with cystic fibrosis. Jacob Nelson, M.D.; Peter Karempelis, M.D.; Jordan Dunitz, M.D.; Ryan Hunter, Ph.D.; Holly Boyer, M.D. International Forum Of Allergy & Rhinology. 6 December 2017

The researchers examined if sinonasal secretions were transferred to the lungs in patients with cystic fibrosis (CF)-related sinus disease and healthy individuals, especially in the recumbent position and during sleep. Evidence was produced regarding the aspiration of sinonasal secretions into the lungs of patients with CF and healthy adults in

the recumbent position. Since both patients and controls aspirated secretions, it was deduced that aspiration alone did not account for the pathogenesis of lung disease in CF patients. <http://tinyurl.com/y9fxj327>

Correlations between cystic fibrosis genotype and sinus disease severity in chronic rhinosinusitis. Waleed M. Abuzeid, MBBS; Changeun Song, M.D.; Judd H. Fastenberg, M.D.; Christina H. Fang, M.D.; Noel Ayoub, BS; Elina Jerschow, M.D.; Paul K. Mohabir, M.D.; Peter H. Hwang, M.D. The Laryngoscope. December 13, 2017

An assessment was carried out of the impact of *cystic fibrosis transmembrane conductance regulator (CFTR)* genotype functional classification on sinonasal disease severity in patients with chronic rhinosinusitis (CRS). No link was determined between high-risk CFTR genotypes with worse sinonasal disease severity or postoperative symptom control than low-risk CFTR genotypes after adjusting for confounding factors. <http://tinyurl.com/ycc35cbx> ▲

Laura Tillman is 70 and has CF. She is a former Director and President of USACFA. She and her husband, Lew, live in Northville, MI.