

## COVID-19 And You: Staying Safe With Current Science And Common Sense

By Xan Nowakowski, PhD, MPH

Let's talk about #Coronavirus, folks. As of March 11, the novel coronavirus first spotted in 2019 has achieved global pandemic status per the World Health Organization.

These are big words about a tiny organism that can hurt people's lungs badly.

The COVID-19 virus is particularly nasty because it has a big spike of protein on it that lets it bond easily to other proteins that help our lungs work. In some people who contract the virus, this sets off a chain reaction where particles called cytokines bombard and damage the lung tissue. The end result for many of these folks is... pulmonary fibrosis.

Proteins not working properly? Wildly aggressive immune activity? Thickening and scarring in the airways? If this sounds familiar, you're thinking along the right lines! And you're probably thinking about how to

“ Keep those hands clean and your risk of getting COVID-19 near your own lungs or a loved one's goes way down. ”

keep yourself safe as the virus continues to spread worldwide.

First and foremost: **It's okay to feel afraid.**

I need to say right from the start that our community should remember the difference between being “susceptible” to harms from a particular virus and actually being “vulnerable” to getting exposed to and infected with that virus. These are concepts from epidemiology—science focused on how infectious and chronic health conditions start and progress. Because of my unique background as a public health scientist and medical educator who lives with CF as a chronic condition and multiple infections as acute challenges, I am very

interested personally and active professionally in both of these areas.

So let me break things down for those who are just getting oriented! Almost everyone with CF is susceptible to COVID-19 because we often deal with very similar damage to our lungs already. By contrast, not everyone with CF is particularly vulnerable to COVID-19. Whether or not we are vulnerable as individuals depends on a few things, but mostly (A) whether or not we have weakened immune systems and (B) what activities we do each day.

I'll get to both of those things in a bit. For now...we should remember these important differences while also

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# Information From The Internet...

Compiled by Laura Tillman

## CF Patients Need Routine Screenings For Iron Deficiency, Study Suggests

Iron deficiency is common among adults with stable cystic fibrosis (CF) and is significantly associated with anemia, vitamin A deficiency, antacids use, and moderate to severe lung disease. Prevalence rates of iron deficiency were found to vary widely due to poor agreement between several laboratory indicators of iron status. Thus, researchers recommended that guidelines be developed for routine and standardized screening, as well as for diagnosis and management of iron deficiency in CF patients. Iron is a micronutrient involved in several important biologic processes, but is mainly associated with



LAURA TILLMAN

the production of hemoglobin, the oxygen-transporting protein present in red blood cells. Iron deficiency is the main cause of anemia (low levels of red blood

cells or hemoglobin), which is then classified as iron-deficiency anemia. Anemia can lead to weakness, shortness of breath, heart problems, and increased risk of infections. Several studies have estimated that 28%–69% of CF patients have iron deficiency. However, the lack of standardized values to classify iron deficiency, and the fact that many of these studies assessed iron levels regardless of CF patients' disease state—which is known to influence iron values—challenges the interpretation and generalization of these data. In addition, despite a seemingly high prevalence rate, few centers are routinely screening for iron deficiency, and there are presently no guidelines on screening, diagnosis, or management of iron deficiency specific to the CF population. Thus, a team of researchers set out to identify the frequency of iron deficiency and anemia in adults with stable CF, and the risk factors associated with both conditions. They also compared iron deficiency

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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. E-mail all submissions to: [cfroundtable@usacfa.org](mailto:cfroundtable@usacfa.org). Or go to our website: [www.cfroundtable.com/newsletter](http://www.cfroundtable.com/newsletter).

**Spring (May) 2020: Weight Issues.** (Current issue.)

**Summer (August) 2020: Diversity in the CF Community.** (Submissions due June 15, 2020.) How do you raise awareness in the CF community? How do you cope with feeling isolated or different within the CF community? What support, research, or recognition would you like to see in the community?

**Autumn (November) 2020: People with CF Who Have Started A Business.** (Submissions due September 15, 2020.) What type of business have you started or are planning to launch? Was it geared towards the CF community or to the general population? What has been helpful? And what have you learned from your experience?

**Winter (February) 2021: Late Diagnosis of CF.** (Submissions due December 15, 2020.)



## ASK THE ATTORNEY

# Pandemic, Workplace Rights, And Cystic Fibrosis<sup>®</sup>

By Beth Sufian

**T**he COVID-19 pandemic has led many employers to have employees work remotely from home as a means of preventing further spread of the virus. Many working people with cystic fibrosis have asked about their employment rights as individuals with disabilities in the context of a pandemic.

The information provided below is not legal advice about a specific situation and is only meant to be legal information. If you have questions about employment accommodation, the Family Medicaid Leave Act, unemployment benefits or short-term disability benefits, please contact the CF Legal Information Hotline at [CFLegal@sufianpassamano.com](mailto:CFLegal@sufianpassamano.com) or call 1-800-622-0385.

### 1. E.E.O.C. Guidelines.

In 2009, during the H1N1 pandemic, the United States E.E.O.C. issued guidance addressing disability civil rights during a pandemic. The Americans with Disabilities Act (“ADA”) and other anti-discrimination laws continue to apply during a period of pandemic. The laws are not magically suspended during a pandemic, nor is the existence of a pandemic a defense to discriminatory treatment in the workplace.

### 2. ADA is relevant to a pandemic in three major ways.

ADA is relevant in times of a pandemic in at least three major ways. First, the ADA governs when and how covered employers may make disability-related inquiries or conduct medical examinations. Under the ADA, only employers with 15 or more employees are covered by the act.

Second, the ADA prohibits covered employers from excluding an individual with a disability from the work-

place, except in instances of a direct threat to the health and safety of the individual or others. “Direct threat” is a legally defined term, which means a significant risk of substantial harm, even with a reasonable accommodation.

Third, the ADA requires covered employers to provide reasonable accommodations to individuals with disabilities, even during a time of pandemic. Employers are excused from providing reasonable accommodations only when the accommodation imposes an undue burden on the employer.

### 3. Disability inquiries and examinations.

Employers are generally prohibited from making disability-related inquiries or conducting medical examinations. A disability-related inquiry is any inquiry that is likely to elicit information about an individual’s disability.

A. A pandemic is not a reason to

make general inquiries about chronic illnesses.

Before a pandemic occurs, employers are not permitted to ask if an employee has a compromised immune system or a chronic health condition. However, if a pandemic occurs and the objective evidence about the pandemic virus indicates that the virus presents a direct threat to individuals with compromised immunity or specific chronic illnesses, then the employer may inquire if a person is at significant risk of substantial harm for the purpose of protecting them from a direct threat.

B. Inquiries when calling in sick or requesting sick leave.

In the absence of a pandemic, employers are not permitted to make disability-related inquiries when an employee calls in sick or requests sick leave. However, during a pandemic, the employer may ask a worker who calls in sick or requests sick leave about whether the sick worker has any symptoms related to the pandemic disease. This is not regarded as a disability-related inquiry because it does not elicit information about the employee’s disability or chronic health condition, if any. Rather, the inquiry is only to the extent that the symptoms are related to the pandemic disease and only to the extent necessary to assess the possibility of a direct threat to the health and safety of other workers.

If, during a pandemic, an employee displays symptoms associated with the pandemic virus while working, the employer may send the worker home or require the worker to remain home on sick leave.

C. Inquiries when an employee returns from travel.

When a worker returns from traveling during a pandemic, an employer



BETH SUFIAN

may ask about his/her possible exposure to the pandemic virus based on the people and places they visited, modes of transportation, and other factors. These are not considered disability-related inquiries. The inquiries are permitted to assess a direct threat to the employee or others. The employer may ask about travel-related exposure to pandemic viruses regardless of whether the purpose of the travel was personal or business. The employee does not need to exhibit any symptoms of the pandemic virus before the employer may ask about possible exposure.

D. Requests to work remotely or adopt safety precautions.

Employers may ask or encourage employees to work remotely from home as an infection control precaution. Employers may require employees to adopt infection control practices and use personal protective equipment.

However, even these pandemic-related, work safety rules are subject to the employer's obligation to provide a reasonable accommodation under the ADA. For example, if the employer requires all employees to wear gloves while working, an employee with a severe latex allergy may request the accommodation of being supplied with latex-free gloves.

#### **4. Reasonable accommodations before and during a pandemic.**

A. Obligation to provide accommodation even during a pandemic.

If an employee is receiving a reasonable accommodation before a pandemic arises, then the employer is required to continue to provide that accommodation during the pandemic unless circumstances change, and the accommodation becomes an undue hardship on the employer.

Similarly, if the need for a reasonable accommodation arises during the pandemic, an individual with a disability may request one, including accommodations that are directly related to

the pandemic conditions. For example, if a worker with CF needed an accommodation to work remotely from home to prevent exposure to the pandemic because of the heightened susceptibility of a person with CF, the employer should grant the request, provided doing so would not impose an undue hardship on the employer.

B. Reassignment of work.

Many jobs, however, cannot be performed remotely and require the worker to be present at the workplace; for example, if an essential function of the job is to work as a server at a restaurant.

An accommodation is a change that allows a person to perform the essential functions of a job. If the essential functions of the job require performance of the job duties in person at the workplace, then it is unlikely there is a reasonable accommodation that can be provided. An employee can ask if there is another vacant position that could be performed at home.

C. Only a disabled employee is entitled to a reasonable accommodation.

Reasonable accommodations, however, are only available to accommodate a disabled individual who is an employee. Consequently, if a non-disabled employee asks to work remotely from home as an accommodation to prevent the employee's spouse with CF from being exposed to the pandemic virus, the employer would not be obligated to grant the request because the employee is not an individual with a disability.

Even if, in this example, the employee requested leave under the Family and Medical Leave Act ("FMLA") to provide care for a spouse with CF, the employee would not achieve the desired goal. FMLA is not the opportunity to work from home—it is a leave of absence from work. The FMLA envisions that leave would be required to care for the spouse. As such, the spouse or parent should pro-

vide proof that the spouse or parent now needs to be at home to assist the person with CF with care and treatment.

In times of a pandemic, however, the employee and employer may simply agree to a new temporary work arrangement for a spouse or parent that permits remote work from home, even if remote work is not an entitlement under the ADA or FMLA.

#### **5. Returning to work after a pandemic.**

After the pandemic ends, an employer may require any employee who has been away from the workplace to provide the employer with certification from a doctor that the employee is able to return to work, provided that all similarly situated employees are treated in the same manner.

#### **6. Other options.**

A person with CF who is unable to work may be able to apply for private short-term disability benefits offered by their employer if the employee cannot perform their job due to the pandemic virus. An employee who has been laid off or terminated from full-time work may be able to apply for unemployment benefit compensation.

People with CF who work part time and also receive Social Security Disability benefits should make sure they understand the risks of receiving unemployment for part-time workers. A person who receives SSI benefits will have the SSI reduced by any unemployment benefit for part-time workers.

Please send any additional questions to the CF Legal Information Hotline. ▲

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*Beth is 54 and has CF. She is an attorney who focuses her law practice on disability law and is the President of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues at [CFLegal@sufianpassano.com](mailto:CFLegal@sufianpassano.com).*



# SPIRIT MEDICINE

## The Spirit of Fear

By Isabel Stenzel Byrnes

I'm writing this article while I'm secluded at home, as we are in the midst of a global pandemic of COVID-19. We in the cystic fibrosis community are among the most vulnerable. Everyone around us is hoarding supplies; the fight for resources has begun. Even healthy, young people are fearing contagion from any social contact. It is an unprecedented time.

The CF community is at the forefront of dealing with cross-infection. In the CFRI community we have learned how to connect with each other without touching, to stay six feet apart, or to meet through virtual platforms. We have learned at an early age to wash our hands properly. For those of us who are transplanted, touching our face became taboo the moment we came out of surgery. We are ahead of the game.

Still, the stories fed from the media and the medical community shed light on the seriousness of this virus. The fear and anxiety are palpable. We don't know what is real and what is exaggerated.

The CF community is no stranger to fear. In this article, I thought I'd ponder on fear and the spiritual practices that help us live with fear. I wrote about fear many years ago, but it's worth exploring again.

So let's look at fear. When we are aware of fear and carefully examine it, we can turn it into *conscious* fear. Conscious fear has less power; it feels like it can be harnessed and contained. Fear is our most primitive animal survival instinct. It is a protective emotion that helps us respond to a real or imagined threat. How we respond to fear is

based on our past experience with trauma, as well as our genetics. Our parents' predisposition to anxiety will influence us. And fear is biological. It releases chemicals that impact our bodies. Our adrenalin and stress hormones will help us fight, freeze, or flee. Sometimes, fear can be so overpowering that we might become numb or avoid what makes us scared.

What can we do with fear? We can do a fear check. We can ask ourselves, "What is really dangerous right now?"

*Sometimes, fear can be so overpowering that we might become numb or avoid what makes us scared.*

I'm on the couch. Food is in the fridge. I have my meds. But I can't go to Costco to get my organic lettuce. I think I'll be ok. Phew! What privilege!

In all seriousness, fear, an emotion, feeds thought, which then feeds behavior. We can examine what can be controlled and what needs to be surrendered. Fear motivates us to do something. Rather than compel us to hysterical hoarding, fear can be channeled into positive actions. We can harness a support team, order our meds, avoid

the ER, do our treatments... the list goes on and on. We can be aware of our surroundings and wear a mask or gloves at the grocery store. We can surrender to our vulnerability and ask our boss to work from home. We can also become

hermits. We can rely on alcohol to take the edge off fear. We can tell ourselves what we can't do and diminish our lives. We can deny risk so much we become reckless. We have to be very mindful of what behaviors could be detrimental to our well-being over the long haul.

For me, I deal with fear in many ways. I take care of myself, and try to optimize my physical fitness by exercising, which seems to ward off pathogens and reduce the stress hormone in my body. I also use my mind. Besides primitive defense mechanisms (denial, projection, rationalization, humor, etc.), I tell myself, "Right now, I'm okay. I'm safe."

I also think of the history of my fellow human beings. I think of European Jews in the 1940s and how Anne Frank's family was held up for



ISABEL STENZEL BYRNES

two years. I think of so many generations impacted by smallpox, typhoid fever, or the plague. If they could endure fear and uncertainty, so can we. We know that people are living today with fear of being killed in war zones or not having food on their plates. If they can endure that, I can endure this social distancing. We humans have an enormous capacity to endure fear.

Let's face it: much of this epidemic fear is around the fear of death. We CFers are the canary in the mine—we are the most sensitive and vulnerable. And we are no strangers to the fear of death. I am a hospice social worker and our agency embraces the truth that death is a natural part of life. We all know we will die someday, but coronavirus seems to remind us that it could be sooner rather than later. We had a hospice patient who asked not to be visited by staff because of fear of the virus! What an opportunity this epidemic is, now, for us all to seize the moment, appreciate the time and health we have, and surrender to the future.

Finally, fear diminishes with this shared collective impact. I have an urge to connect with close friends and family by phone, text, and online; to hear reassurance and talk through the fear. In this way, the power of fear is reduced.

What are some spiritual practices to help us overcome fear? Spiritual teachings from all faith traditions teach us to face fear and overcome it.

Fear reminds us that we can recognize safety and security as true illusions. To truly overcome fear on a spiritual level, we must abandon the idea of a safe refuge. Even Trikafta gives us the illusion we will live forever. The spiritual truth is all of life is temporary. This means surrendering to the finiteness of our lives, to our illusion of control.

Fear can be an opportunity to examine our worst imaginings and unexamined beliefs. I have a grief

counseling client whose very true fear was that when her loved one died, she would curl into a ball and die alongside them. But then she had an epiphany: "I finally realized that I am going to be ok." Though life was very, very hard without that person, the worst did not happen. Another belief is that death is the end of us, and to die is unimaginably tragic. True, it will be very sad to leave this earth, but our lives continue on in the impact we have on others. We will be remembered in small and big ways. And some of us believe our spiritual lives continue in another place. Death is the end of physical suffering. I don't want to minimize this difficult reality of death, but sometimes our beliefs make death worse than it really is.

Fear doesn't always feel good, so another spiritual practice is deliberately turning toward what does. Fear helps us *decide* to focus on thinking about things that are true, noble, right, pure, lovely, admirable, and praiseworthy. We have choices over our thoughts. These larger truths about the beauty of the world, the love we feel for others, and the essential goodness and wholeness of life get obscured with fear.

We see in the media that fear brings out the best and worst in humans. We have people selling a mask for \$40, but we also see people offering to help people who are shut in. How can our fear be used as a conduit of love, care, and support? Turning to altruistic service is a byproduct of fear...it's *doing* something with our restless, scary energy. And it is a spiritual practice.

Another spiritual practice is trust. To trust that the trials we have offer something to us. What does God want us to do given these circumstances? Live life to the fullest. Face life with joy and love, not fear. Help others less fortunate. Focus on what matters most. Never give up.

Times of fear give new meaning to prayer. Prayer is a form of giving up fear and giving it to God or your own version of a higher being. Prayer means holding onto hope that things will be okay, that things will get better.

So my prayer, or hope, is that by the time this article is published, this wave of hysteria has died down and our fears are placated by a warm, healthier summer coming up. Realistically, some people will be deeply impacted by this virus and fears will be realized. My heart goes out to those whose lives are uprooted financially or who have lost a loved one. Life is unfair. But it is still good. And life is stronger than a virus. ▲

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*Isabel Stenzel Byrnes is 48 and has CF. She lives in Redwood City, California, and is happily married to Andrew Byrnes. She loves to hike, play the bagpipes, and be a slow triathlete. Her email is isabear27@hotmail.com.*

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## *In Memory*

**Peter McMurry, 51**  
New York City, NY  
Died on December 12,  
2019

*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



# SPEEDING PAST 50

## I'm Rooting For Us

By Kathy Russell

I think we have made it through another winter. We have had weather in the high 60s and blue, sunny skies. Then we got a few days of snow flurries. I guess that winter just wasn't willing to go out quietly. My arthritis decided to kick up again, too. At any rate, spring is almost here. Wahoo!

To some that is no big deal, but it is a really big deal to me. Every winter I try very hard to stay away from people who may have any kind of bacterial or viral infection that they are only too happy to share with me. Often, they don't mean to share their germs, but it seems to happen, oh, so easily.

Now with the worries about COVID-19, I am trying to stay away from people in general. Let's hope that we all are able to avoid this new threat. It seems to be getting worse by the hour. Many people are dying from it and most of them are old geezers like me. As Gilda Radner said, "It's always something."

The World Health Organization ("WHO") and the Centers for Disease Control and Prevention ("CDC") are putting out guidelines for how we should be acting with this threat. I find it very interesting that the same people who are telling us how we should behave during this medical crisis are acting as if they feel immune to it. I saw the President, heads of several large corporations, and several government officials together at a press briefing from the White House where they were all shaking each other's hands, standing side-by-side and all using the same microphone. They also touched their faces after

touching the mic. One even coughed without covering his mouth! These are the very people who are telling us what we "must" do to protect ourselves and others.

Actually, these guidelines are probably easier for those of us in the CF community than they are for the average person. We are more accustomed to keeping adequate distance between ourselves and others, although I am not used to keeping 10 feet from oth-

ers, which is the newest guideline that I have heard. Also, we are used to using hand sanitizer or wearing gloves, wearing masks, and not shaking hands. My pulmonologist and I have used elbow touches for years. Now the recommendation is to not touch at all but to use a hand held over the heart to signify a greeting. I can do that.

My husband and I are separating ourselves from others. Since we live in our own home, which is set on a couple of acres, we needn't worry about getting too close to neighbors. When I did go out to the grocery store, I wore gloves and a mask. I didn't touch anything but the food I was buying and I washed all of the produce when I got it home. Of course, I always wash all produce and I always wash my hands after returning home.

This pandemic is giving a lot of people a small taste of some of what we, who have CF, have been going through for years. I will say that I haven't heard too much grumbling from most of the regular folks who are being affected by this awful virus. Even in the terribly crowded grocery stores, people stood in long lines without complaining...too much. I imagine that most people are waiting anxiously for all of this to be over so that they can go back to their regular ways of living. We who have CF will still have to keep our distance from others, use hand sanitizer or gloves, and sometimes wear masks. Oh, well, such is life when you have CF.

The various warnings and dire predictions have caused my memory to dredge up times in my youth when we

*"This pandemic is giving a lot of people a small taste of some of what we, who have CF, have been going through for years."*



KATHY RUSSELL

would see quarantine signs posted on people's home. I think that mostly they were for tuberculosis (TB), diphtheria, scarlet fever, and even polio. The Health Department would tack a sign on the front door of homes where someone was infected with a dire disease. No one could go in or out. I remember how scary it was to see these notices (some were bright pink and others had a red banner on them) on people's doors as we drove down a street. Even though I was quite young, it made a lasting, frightening impression on me. I thank goodness that we don't see those signs anymore.

Anyway, I have very sad memories of that time. I was even hospitalized in the City Isolation Hospital because someone, when looking at my chest x-ray, decided that I must have TB. My parents were told that they had to have me at that hospital by a certain time or a "bench warrant" for their arrest would be issued.

The hospital was old, dreary, and had heavy iron bars on the windows. I was 12 years old and was stuck in a large, cold room with nothing but those iron bars to look at. It was quite awful and I have worked very hard to forget that time. This has brought it all up again. I guess it is a form of PTSD. This, too, shall pass.

I am hoping that COVID-19 will be controlled or will run out of steam. I am hoping that all the countries that are affected by it can get a handle on it and that fewer, rather than more, people will contract it. I'm rooting for the people of the world.

Stay healthy and happy,  
Kathy ▲

*Kathy is 75 and has CF. She and her husband, Paul, live in Gresham, OR. She is a former President and Treasurer of USACFA and served as Managing Editor of CF Roundtable for many years. You may contact her at [krussell@usacfa.org](mailto:krussell@usacfa.org).*

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## DIAMOND SUSTAINING PARTNERS





# FAMILY MATTERS

## No Longer “The Skinny One:” Dealing With A Changed Body After Pregnancy

By Cindy Baldwin

If you're a woman who is pregnant or has had a child and are talking with other people who are pregnant or have had children, it's nearly impossible to avoid the topic of weight. "I'm still trying to lose the baby weight." "My doctor thinks I'm gaining too much weight for the first trimester." "My OB is worried about the baby's weight and wants to schedule an induction." Even years after birth, these weight-focused conversations can continue—woman after woman anxiously trying to come to terms with the way her body has been changed by the gift of giving life.

For CFers, weight is a minefield in a different way. Most of us have grown up as “the skinny one,” putting all our focus on the need to gain and maintain weight, never to lose it. Nearly all of us are all too familiar with weight-gain shakes and ways to pack extra calories into each meal; many of us have to turn, at one point or another, to invasive measures like feeding tubes in order to stay at a safe-weight threshold. Weight gain during pregnancy and breastfeeding can be especially difficult for CFers, struggling as we are to hold on to precious nutrients when our baby is efficiently funneling them all away. Many of us have to fight for every calorie, working hard to keep both our baby's and our own weight up.

And so it comes as a surprise to a lot of CF women, that pregnancy can change the topography of our bodies just as much as it does for anyone else.

Unlike many CF mamas, I gained a

lot of weight during my pregnancy—mostly due to the fact that I had polyhydramnios (very high amniotic fluid) and a genetic predisposition on both sides of the family to large babies (my daughter ended up weighing a whopping 9 lb. 8 oz. at birth). Through my pregnancy, I worried occasionally about what it would be like after I gave birth, but I mostly focused on seeing that weight gain as a blessing and went on with life. I was extremely sick during and after giving birth, and the preg-

nancy weight fell away with almost dizzying speed: In less than two weeks, I had lost so much weight I was already under my pre-pregnancy poundage, and I spent the next several months forcing myself to eat a high-calorie snack every single time I nursed my baby so that I could stop the downward spiral of the numbers on the scale.

So I did not expect, in those months after birth, just how *different* my body was going to be from now on. Even with such dramatic weight loss, carrying my daughter inside me for nine months had permanently shifted the way my body looked. Suddenly my lifelong “CF belly” was compounded by the way pregnancy had changed me; what weight I did have sat across my middle in ways it never had before. My mild-but-persistent abdominal separation didn't help, and

neither did the web of stretch marks or loose skin that proved just how much fluid and baby I'd once carried within me. My breasts hung heavier; my hips were wider. I somehow went up a whole pant size, even though my weight was lower than it had been for years.

As my baby grew into a toddler, I sometimes found myself standing in my bedroom in the mornings as I got dressed, staring at the body that had become so unrecognizable to me, feeling as though it must belong to somebody else. I realized, in those postpartum months, how much I'd unconsciously come to define myself by those words people had applied to me all my life: *the skinny one*. My childhood had been one round of Scandishakes after Carnation Instant Breakfast Drinks

*“I realized, in those postpartum months, how much I'd unconsciously come to define myself by those words people had applied to me all my life: the skinny one.”*



CINDY BALDWIN

after another. During puberty, my mom had tacked a chart to the fridge and made me record how much protein and fat I was getting each day, so I could stand the best chance of capitalizing on the growth spurts of adolescence. In high school, friends had always described me with terms like *petite* and *tiny* and *small*.

Even though I'd insisted, growing up, that I just wanted to gain weight and stay healthy, I began to recognize after my baby was born the secret, smug pride I'd taken in my body's shape: the way I could fit into the smaller jeans, wear the tighter shirts, not have to worry about whether my buttons would close. And I found myself wondering: If pregnancy has changed the way I fit into clothes so very much, how will I now define myself?

Many CF patients continue to struggle with severe weight issues into adulthood, and that's an enormous challenge that has been discussed and written about often in CF spaces. But as I've gotten older, and especially as more and more of the CF population has become eligible for CFTR modulators and more CF women have had successful pregnancies, I've found myself in an increasing number of private conversations with adult CF women who, like me, find themselves not recognizing their bodies anymore. Sometimes it's because better medications have led to a little pudgy, or because pregnancy has rearranged things on their still-thin frames in ways that don't align quite so well with the conventional standard of beauty, or simply because age has a way of consolidating any fat we *do* have in the places we least like.

In each of these conversations, the same themes have emerged. "I don't recognize my body." "I don't feel comfortable in my skin anymore." "My clothes fit wrong." And in so many of

these discussions, one thought has lurked under the surface, one most of us are afraid to speak out loud: "Who am I if I'm no longer *the skinny one*?"

These days, my daughter is seven years old and I am not planning to have another child. Even still, my body has never gone back to what it was before I got pregnant. Over time, I've made my peace with that, getting to know my new body, with its curves and squishy places, the spiderwebbing of stretch marks, the c-section scar. Two years ago, tired of the way fly-front jeans always put pressure on my perpetually-tender abdomen no matter what size I wore, I decided to make a change: instead of fighting the body I had and wishing for one I didn't, I was going to use the way I dressed to cherish myself. I wanted to worry more about what *feels* good and less about what *looks* good. So I put my old jeans in storage and replaced them with pants that didn't feel like torture to wear; these days, I have a whole wardrobe of stylish leggings, professional-looking slacks you'd never guess were really yoga pants in disguise, and soft dresses that hug my body without pain and without judgment.

I realized, this winter, that I feel beautiful again, comfortable in the person I've become.

If you're a person who's pregnant or had a child, you may know all too well the feeling of looking down at yourself and thinking, *I do not recognize this body*. But from one cyster to another: you are beautiful, mama. Skinny, soft, stretchy, pudgy, or lumpy, that body you have? It's worth celebrating. ▲

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*Cindy is 31, has CF, and lives just outside Portland, Oregon, with her husband and seven-year-old daughter. She is the author of Where the Watermelons Grow, Beginners Welcome, and the forthcoming The Stars of Whistling Ridge, all with HarperCollins Children's Books.*



## What is the Boomer Esiason Foundation?

In 1993, NFL Quarterback, Boomer Esiason, learned that his son, Gunnar, was diagnosed with the incurable genetic disease cystic fibrosis (CF). Never ones to back down from a fight, he and his wife, Cheryl, founded BEF and decided then and there to fight for a cure and for the cystic fibrosis community.

Cystic Fibrosis is an inherited chronic disease that affects the lungs, digestive system, and reproductive system of about 30,000 Americans by causing a thick build-up of mucus that leads to blockage, inflammation, and infection.

## What does BEF do?

In addition to assisting the CF community with the following programs, we also support CF clinics and research centers:

- Educational Scholarships
- Lung Transplant Grant Program
- Team Boomer
- Jerry Cahill's Cystic Fibrosis Podcasts & Wind Sprints
- Breathe In Podcast
- CF Patient Disaster Relief Program
- CF Step-by-Step Video Series
- Gunnar Esiason Blog
- Tru Heroes Nursing Program
- You Cannot Fail Hospital Bags
- CF Education Days & CF Speaking Engagements

[www.esiason.org](http://www.esiason.org)



## CFRD EDUCATION

# The Impact Of Diabetes On Weight Management

By Kat Porco

When we talk about CF, the recurring conversations generally revolve around pulmonary health and the hurdles associated with weight management. We know that weight loss can be associated with a number of issues: disease progression, poor pancreatic function, or a side effect of either untreated or poorly treated diabetes. As testing and vigilance around diabetes are on the rise, we are starting to see more and more diagnoses being made. Over 50% of CF patients over the age of 30 are estimated to have CFRD, which many endocrinologists believe is highly underestimated due to poor follow through on oral glucose tolerance testing.

Diabetes can potentially exacerbate not only the pulmonary status, but also inhibit an individual's efforts toward weight gain. We often see a decline in weight up to three years prior to a diabetes diagnosis in the CF community. To understand why that happens, you have to understand the role of insulin, a hormone, in the body and the cellular processes involved. When food is digested, glucose is released into the bloodstream and the pancreas, in response, releases insulin. Much like a lock and key, the insulin attaches to receptors at the cell surface, signaling another molecule to transport the glucose into the cell, where it can be used for energy. When insulin is not available, either due to deficiency or resistance to insulin, glucose cannot enter the cells and be used for energy. This insulin deficiency, whether

endogenous (insulin released from the pancreas) or exogenous (injected insulin), causes a buildup of glucose in the blood, ultimately resulting in high blood sugars. When the body cannot utilize the glucose, it, in turn, believes that it is starving and develops a work around to compensate for the lack of energy available. In these situations, the body can fall back on burning fat and muscle. However, when this happens, the body loses fat and muscle at a very rapid rate, resulting in weight loss.

So, if you have noticed unexplained weight loss over the past few years and have not yet been diagnosed

with CFRD, it's advisable to have your sugars checked and oral glucose tolerance test administered to confirm that you are not diabetic. Untreated diabetes is not only difficult on your pancreas, causing it even more damage, but it also creates an ongoing struggle for your kidneys as they are trying (often unsuccessfully) to rid the body of the excess sugars present when insulin is not there to do the job.

Admittedly, diabetes throws an additional wrench in the CF mix, as we have not only the day-to-day concerns of pulmonary therapies, but we now have the chaos and hurdles of diabetes to balance as well. However,

treating diabetes is critical for the most ideal pulmonary and vascular health outcomes. ▲

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*We often see a decline in weight up to three years prior to a diabetes diagnosis in the CF community.*

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*Kat Porco is a diabetes educator and the co-founder of Attain Health Foundation.*

*She received a Bachelor's in*

*Social Work followed by a Master's of Science in Health Communications. Her work over the past ten years has been solely focused on supporting and advocating for the cystic fibrosis community. Throughout these years, she has seen the disconnect between the recommendations of the medical community and applicability for the patient community. Because she does understand this complex relationship, Kat felt that she could assist in bridging the gap to reach ultimate health goals through health coaching and diabetes education. She is a Duke Certified Integrative Health Coach, as well as being Nationally Board Certified in Health & Wellness Coaching through the National Board of Medical Examiners. Kat is a Certified Diabetes Care and Education Specialist (CDCES) through NCBDE.*



**KAT PORCO**



# ASK THE ENT

## Your ENT Questions Answered

By David Gudis, MD

NEW  
COLUMN

**H**ello CF Roundtable Readers! Please allow me to introduce myself. I am David Gudis, an Ear, Nose, and Throat doctor at Columbia University Irving Medical Center in New York City. I am very interested in the care and management of patients with CF sinus problems and one of my patients told me about this fantastic newsletter. Almost every CF patient will have an ENT problem at some point. Most ENTs around the country do not see many CF patients, so it can be hard to find the right answer. Therefore, I thought we could create a great resource here for the CF Roundtable Community. Going forward, we invite you to send any of your CF ENT questions (e.g., Why am I always a little hoarse? Why is my sense of smell diminished?) to: [croundtable@usacfa.org](mailto:croundtable@usacfa.org). We will pick a few from the list for each “Ask the ENT” Column!

A little about myself: After completing my residency in otolaryngology (head and neck surgery), I completed two fellowships, or sub-specialty training: one in pediatric otolaryngology, and one in rhinology—sinus and skull base surgery. Most of my practice involves complex endoscopic sinus and skull base surgery. CF chronic sinusitis is a medical problem that bridges these two sub-specialty interests of mine. I am very fortunate to practice at Columbia University, where our pulmonologists, pediatricians, and transplant doctors have built an outstanding cystic fibrosis center. And the simple truth is that CF patients are the strongest, kindest, and most resilient patients I’ve met. So, please let me know your questions—I look forward to hearing from you!

**Q: How do seasonal allergies**

**impact treatment of CF sinusitis? How do you recommend allergies are tested for and treated?**

A: About one-third of CF patients also have allergic rhinitis. The symptoms of chronic rhinosinusitis (“CRS”) and allergic rhinitis (“AR”) can often overlap. Although both conditions can cause nasal congestion and nasal discharge, there are some important differences. Nasal discharge from AR is typically clear and watery and generally does not cause sinus pressure or pain. AR is often associated with sneezing, itchy eyes, and an itchy nose. CRS, on the other hand, causes thick, discolored nasal discharge and may be associated with pain or pressure around the eyes, cheeks, or forehead. The best treatment to try first for AR is a nasal steroid spray, like fluticasone, often in combination with a topical nasal antihistamine spray, like azelastine. An oral antihistamine (like Claritin or Allegra) can help, too. Most patients will have significant



DAVID GUDIS, MD

improvement from those medications making allergy testing unnecessary. If those treatments do not help, then allergy testing may be useful so that your allergist may consider immunotherapy such as allergy shots or sublingual treatments. Be sure to talk to your doctor before starting any new medications, including over-the-counter medications.

**Q: What are options for treating polyps, especially if caught early enough, to avoid sinus surgery? What are your recommendations for preventing polyp growth?**

A: Polyps are little benign growths in the sinuses that are essentially swollen areas of the mucus membrane lining the nose and sinuses. In CF, polyps are tricky. For most non-CF patients with nasal polyps, the polyps are mostly from eosinophilic inflammation, which means they respond well to steroids. Nasal steroid sprays, or prednisone when needed, can shrink the polyps. In CF, the polyps are unfortunately from mostly neutrophilic inflammation, and are not as responsive to steroids. Therefore, nasal steroid sprays and prednisone generally do not work as well on polyps in CF patients as they do in non-CF patients. However, there is one study from several years ago that did demonstrate some reduction in polyp size from nasal steroid spray. In addition, most patients have some mix of “eosinophilic” and “neutrophilic” inflammation. Therefore, since side effects are so rare and mild, it is almost always worth trying nasal steroid sprays for CF patients with polyps. Surgery remains the most effective way to treat polyps in CF. ▲

acknowledging full stop that a global pandemic is a scary situation. COVID-19 may feel particularly frightening within our community because of its potential for harmful impacts on the lungs.

It's also not remotely unusual for people in our community to experience those kinds of fears—whether during a global pandemic or a shopping trip. Because of this, we are both acutely aware of the stakes here and uniquely equipped to help everyone stay safe. In a world where countless others are waking up to the importance of conscientious and consistent infection control, our small but mighty patient community is full of superheroes. If you've ever thought Big Air Jerry has the coolest job in the world, now's your chance to shine!

So how do you become an infection control superhero while making sure to put on your own oxygen mask first? That actually begins with remembering that masks aren't a very powerful weapon against most viruses!

People with CF are accustomed to masking appropriately in circumstances where it helps. You absolutely want to keep up with your usual routines, including any protective equipment your team recommends. But you can also help others understand why you need to wear masks in certain situations. Spreading accurate information about when masks actually help—and what kinds should be worn in those very specific circumstances—ensures protective supplies are consistently available for those needing them.

The good news here is that masks don't help much because COVID-19 is not airborne in and of itself. It spreads through inhalation of small droplets of saliva and mucus, either directly from another person's mouth or—much more commonly—left behind on surfaces that lots of people touch. Yes, some people's mucus is actually liquid! But even when it is, it can't fly very far. The biggest risk to people with and without CF alike is touching contaminated surfaces, then touching your mouth or nose and get-

ting those organisms into your lungs. Although a mask may reduce such hand contact, it can also create a false sense of security and limit handwashing before removing or adjusting the mask. There are conflicting recommendations from the CDC and WHO right now about masking. The most important thing to remember if you do wear a mask when running essential errands is to put on and remove it using proper technique, and to continue following all other recommended protective measures.

Consequently, the even better news is that COVID-19 is easy to kill. As an



**XAN NOWAKOWSKI**

infection control superhero, your absolute best weapons in this fight are good old-fashioned soap and water for your hands, and basic surface cleaning supplies for everything else. Of course, you already knew that much—you are reading *CF Roundtable*, after all! So you likely practice frequent handwashing and cleaning and set an example for others in your community of how to clean vigorously but sensibly. You can find comfort in knowing that these really are the absolute best ways to make sure you don't breathe in those nasty viruses that can hurt your or a loved one's lungs.

I do need to offer a caveat about what protection approaches work best for each person with CF: our commu-

nity is diverse, and some of that diversity comes from organ transplants. Some of us have made it well into adulthood with all our original major organs. I've lost most of my tooth material and had all my gum tissue surgically reconstructed, but so far am forging ahead with the same lungs. That is not true for everyone, and there are special considerations for people with transplants.

Whether it's your lungs or other organs that are donated, it's important to follow your usual infection control routines in consultation with your care team. Even if you fall into that transplant group, available evidence still clearly indicates that aggressive hand and surface hygiene are still the best way to protect yourself from exposure to COVID-19! And if you're not transplanted or otherwise living with weak immunity, your risk for developing serious complications if you are exposed is substantially lower.

Did I mention it's super important to wash your hands? Thirty seconds with soap and water, every time. Try humming the chorus of a song you like if you feel the urge to rush. Keep those hands clean and your risk of getting COVID-19 near your own lungs or a loved one's goes way down. So this is the most helpful thing we can all do, regardless of whether we have CF ourselves, to "flatten the curve" of this pandemic.

If flattening the curve is a new idea for you, think of yourself as a novice mountain climber who is just learning the basics. Now imagine two mountains: one that is very tall and narrow and pointy, and one that is very short and wide and smooth. Which one will you try to climb? Well, that gentle little hill you feel ready to start out with is about what our health systems are currently ready for, too.

Slowing down the spread of COVID-19 as much as possible helps avoid a situation where our healthcare facilities and providers are so overwhelmed that they can barely function. This also ensures that people needing supportive care due

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to serious complications can get it promptly when that time comes. And basic hand and surface hygiene are the absolute best ways to get us climbing that easy little hill as we continue to build our skills in treating and eventually vaccinating for this coronavirus.

That said, there's also plenty of other stuff you can do to help keep yourself and others as healthy as possible as the pandemic continues. So, for folks who are newer to following CF infection control guidelines or looking to be more adherent, here are some other simple and thoroughly evidence-based strategies to be an infection control superhero, now and every day!

#### **1. Practice social distancing.**

What this looks like can vary from person to person, but the basic idea is always the same: don't come into close contact with other people any time you don't need to. This is why you see big events and school classes getting canceled, aggressive contact precautions imposed for public spaces, and more people working from home or discouraging office visitors. It's a numbers game where you try to minimize how many people could potentially get exposed at once, while also allowing time and space to clean surfaces and restock handwashing supplies. But it works!

**2. Avoid unnecessary travel.** This is a definitely-not-kissing cousin to social distancing that dramatically slows the spread of viruses to new communities. Think about making a glass of iced tea using a powdered mix. Even if you pour lots of mix into the glass before adding water, it doesn't get dispersed until you stir! So keep the mixing to a minimum by only traveling outside your usual spots when necessary. And if you must travel, take your usual precautions to minimize infection risk.

**3. Buy needed supplies mindfully.** If you or a loved one have CF, this may be easy because you likely have a substantial stash of basic supplies that you use every day to protect yourself and

keep surfaces clean. But remember that, for the most part, your usual routines are going to be as effective in this scenario as any. If this includes wearing masks because of your specific circumstances, great. But don't start masking if you saw no reason to do so before this pandemic gained steam. It's likely to hurt more than it helps unless your clinicians have suggested it. Otherwise, stick to hand and surface cleaning supplies. Replace what you use, but don't hoard.

**4. Cover your hands when touching common surfaces—or just don't use them!** Note that this does not say "wear gloves." If you have CF and are wearing single-use gloves for high-risk activities like pumping gasoline, continue. Otherwise, leave the gloves for chronic illness patients and health professionals, and instead use common sense to avoid touching your mouth or nose with dirty hands. If you're riding elevators, cover your finger with your sleeve before hitting any buttons—or better yet, use your elbow! Do the same when opening doors—or better yet, use your hip! A tissue or paper towel usually works fine, too. Or what about a cardboard drink sleeve, or a used cleaning wipe? Anything is better than nothing, provided you still wash your hands regularly.

**5. Remember that sharing isn't always caring.** Being generous with things we enjoy, like favorite foods and beverages, is wonderful. It's also unfortunately very dangerous when we are concerned about spreading a virus. So as you keep your hands to yourself, do the same with your lunch! Choose takeout or delivery service for any meal you or others in your household do not prepare, and do not share food or drinks with anyone except intimate partners. Rule of freshly washed thumb: If you wouldn't kiss someone on the mouth, don't share food or drinks with them!

**6. Share information only from verifiable sources.** Most of us have probably seen lots of dubious and even conflicting reports about COVID-19 fly-

ing around on social media without attribution or links to scientific resources. When you see one of these posts, step away from the share button...and maybe wash your hands while you're at it! Please do share information on staying safe during the coronavirus pandemic on social media. But get your information from appropriate sources like the World Health Organization and national organizations with similar focus. This includes CF advocacy and science organizations for your country, which are great places to look for specialized guidance. If you can't verify the source of someone's post, ask them! They may delete the post if you point out the danger in sharing unverified information.

**7. Get enough sleep.** I fully admit to struggling with this myself, and I suspect I'm not alone. Heightened awareness of how scary and dangerous the world is for our community can make it tough to fall asleep and wake rested. I don't find this part as difficult, but your essential-travel mileage may vary! What I find harder is not answering every person's questions or calming every fear I see in my networks. I take the superhero thing a bit too seriously sometimes! But I'm trying to do better. After all, my work reminds me every day how important adequate sleep is for preventing CF complications and other problems before they start. So after you've counted to 30 while washing your hands, take a break and count some sheep instead. This will help you resist infection if you are exposed to COVID-19, which makes you and everyone else much safer.

**8. Stay nourished and hydrated.** If you have CF, now's your chance to recreate silly memes from your CF social media groups by eating nutritionally dense food as needed. You may not need as many calories anymore if you use a modulator drug like Trikafta or Kalydeco! But folks not on modulators must meet our energy requirements even more conscientiously. And we always

Continued on page 19



# BREAKING NEWS

## COVID-19 PANDEMIC AND CF

# COVID-19 In Italy

By Francesco Longo

**Q: Did you have time to prepare for the quarantine? If so, what did you do in preparation?**

A: The short answer is no, we were not warned and couldn't prepare. The situation evolved very rapidly, there were no warnings, and at times it felt like no one knew what was going to happen on the next day.

The first containment measure was issued on the 23rd of February, and it involved a small area of 11 towns in the Lombardy and Veneto regions, as well as any individual who had been there in the previous 14 days. At that time, we were told that this would effectively prevent the spread, but the first signs of what was coming were already there. I remember trying to source a Cambridge mask, only to find out that they were out of stock. In fact, since then I've been unable to source a personal protection mask at all.

As contagion kept rising and spreading geographically, the containment measures grew across northern Italy, both in area coverage and in the intensity of restrictions, but it felt like the virus had a head start on us. Lombardy and a few more provinces were placed under lockdown on the 8th of March, but this information was leaked by the press the night before, and thousands of people left immediately for other regions of Italy and other countries. Soon after, on the 10th of March, the whole country was officially involved in the containment measures, which were also ramped up to the highest level.

**Q: How long have you been in quarantine?**

A: Formally, where I live in the Turin region, we've only been involved

in the quarantine since the 10th of March, when the whole country was included. However, given that we live a mere 150km from the contagion hotspot, and that there were several confirmed cases at hospitals as close as 5km from our home, we effectively self-quarantined since late February to stay safe.

**Q: Are you still able to get all of your meds during the quarantine? If so, how are you getting them?**

A: We don't know what's going to happen in regard to this. I've already started to have some issues, as I'm running out of remote blood glucose sensors and there's no estimated delivery date for replacements. As for the standard CF meds, from pancreatic enzymes to hypertonic saline and the like, I will need to restock in a month from now, as we are on a two-month restock cycle, and so far we've been told that access to drugs will be guaranteed, but I feel that this guarantee is more geared towards the general public and that



FRANCESCO LONGO

there may be supply issues for specialty medications.

**Q: How are you sourcing your food and supplies?**

A: Despite the news speaking of quarantine, it's not exactly that. It's more akin to an economic and social lockdown. People can still leave the house for these reasons: to buy food and essential necessities; to buy drugs; to commute to work; to provide assistance to relatives; and for medical emergencies. Currently, only supermarkets, pharmacies, some convenience stores, and other essential public services, as well as manufacturing plants, are operating, with few exceptions.

**Q: What is happening when people with CF need care and what does the reallocation of CF healthcare providers look like? Are you able to get in touch with your doctors in the event of an emergency?**

A: Since late February, all my regular appointments were postponed indefinitely. I attend both a CF clinic attached to one hospital, and a transplant center attached to another, as I'm on an active transplant waiting list, and both hospitals are now fully involved in COVID-19 management. In Lombardy, the hardest hit region, I've been told that the CF ward has been re-designated to house COVID-19 patients, since they're desperate for beds and equipment.

This goes beyond CF, as oncology patients, immunosuppressed patients, and all other patients with chronic healthcare needs are now told to not go to the hospital unless absolutely necessary, to stay at home, and wait until the situation is back to normal. Unfortunately, it seems that risk groups like us are not a priority amidst a healthcare crisis.

**Q: What are your plans if you need**

**care? Are virtual clinic visits an option? What has your CF center told you about seeking care and are they communicating with you regularly about your care needs and options going forward?**

A: I am doing my very best to remain stable, because I wouldn't know what to do if I got sick. In the event of a medical emergency, we would have to visit the ER as usual, but all ERs are now involved with COVID-19 patients and there's risk of contagion. They have not set up parallel, safe access for chronically ill patients. I used to be a very "high-maintenance" patient; I've been waiting for a combined transplant for a long time now; and I have several CF-related comorbidities such as advanced liver disease, multi-resistant bacterial colonies, diabetes...you can imagine. To manage all that, I was having at least weekly checkups, sometimes twice a week, but now it's been three weeks since the last one, and it seems that it'll be at least another three weeks until the situation starts to improve and hospitals can go back to a more normal schedule. In case of need I've been told to call beforehand, if possible, but I fear that I might have a critical situation since it's happened before and find an overflowing ER with overworked, tired staff.

**Q: How are the isolation protocols for CFers different, if at all, compared to other people in isolation? Are you completely isolated from others or are friends and family visiting you in your home?**

A: The government hasn't issued any formal guidelines for CF patients or any other risk groups beyond the recommendation to stay at home. Our patients' association and some individual clinics are recommending the standard levels of precautions for CF patients: washing hands regularly and avoiding contact with people who might have been exposed to the virus.

On top of that, there's the strong recommendation to stay at home, and to avoid hospitals unless absolutely necessary. Then again, now that everything is shut down, we should be safe.

Before that, many patients were still going to work until the national lockdown and, unfortunately, some working in supermarkets or manufacturing still are. There hasn't been any mandatory work leave for risk groups—it's up to general practitioners to decide on a case-by-case basis if they certify the need for a preemptive work leave or not.

**Q: Are transplant and other surgeries still occurring? Are there any plans for handling these scenarios in the event the quarantine lasts longer than a few weeks?**

A: By decree, all non-urgent surgeries have been suspended since they are redirecting operating room staff and equipment to the intensive care units instead. In some hospitals, ICUs are working at full capacity and above, which is an unsustainable situation for a hospital. The worst affected hospitals have had to transfer non-COVID-19 patients to other facilities.

There hasn't been any formal communication involving transplants so far, other than donors will be tested for Coronavirus, but from speaking with other patients in waiting lists, it seems that they are not happening at all now. Unfortunately, in Italy, the vast majority of lung transplants, almost 95% of them last year in fact, are performed in the northern part of the country, and many of the hospitals that usually perform the most transplants every year are in the most affected area. Since every year in Italy only about half the patients waiting for a lung transplant will receive it, this healthcare crisis could mean a stop of one or two months, which is very significant overall.

As grim as it is, I wanted to add that personally, I'm not living much differently than before because it was

already flu season and I was staying at home most of the time. Many other patients are having a harder time, but they understand the risks and the need for precautions.

The worst affected from this situation seem to be healthy people, as they both don't understand the dire implications of a healthcare system operating over capacity for risk groups like us, and many young people who have said that they don't worry too much about this virus as they think they'll survive it just fine. In that regard, patient 1 is a 38-year-old male who used to compete in multiple sports and work out regularly, as healthy as they come. If anything, someone like him should've barely been affected, and yet he spent nearly three weeks on mechanical ventilation. Doctors are also warning that, as the disease progresses, younger people will start getting sicker and come in for ICU admission—it seems that the elderly were affected earlier. Everyone must pull together and help contain the spread of this pathogen—no one is immune—and while not everyone may fear for himself, everyone knows someone else who may be in great danger if exposed. There's only so much the government can do without people's cooperation. We need to fight misinformation and raise awareness. I appreciate the opportunity to share my experience with the CF community at large, and good luck to us all. ▲

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*Francesco is 27 and has CF. He lives in Turin, Piedmont, and mostly spends his time at a PC, whether it's playing video games, watching movies and series, or listening to podcasts and music. He's been on an active transplant list for close to two and a half years.*

For an update on Francesco's experience living in Italy, head over to our blog: <https://www.cfoundtable.com/blog-1!>



## FOCUS TOPIC: WEIGHT ISSUES

# Body Image: The Perception And Impact

By Dr. Leigh Ann Bray

In my work as a bedside nurse on the Pulmonary Care Unit, I often asked patients, “How did your pulmonary function test go?” Nursing care is often based on changes in numbers, whether it be pulmonary function, body mass index (BMI), weight, etc. Through the interactions and rapport with my patients, I quickly learned that the *impact* of those numbers was the missing piece to the puzzle and often went unnoticed, despite being a concern to the patients themselves. I then made it my personal mission to go back to school, become a nurse researcher, and help improve the quality of life behind those numbers.

My dissertation focused on the differences in various aspects of health-related quality of life between men and women who had CF, including both body image and weight. Through my dissertation research, I learned not only that body image and weight were prominent concerns of those with CF, but that the experiences with both were different between men and women. There was no “one size fits all” approach, and what we did for men might not be as helpful for women. A survey of 123 adults (57 men and 66 women), indicated that men reported a slightly lower or poorer health-related quality of life (HRQoL) in the area of body image and a significantly lower or poorer HRQoL in the area of weight. While a deep dive into these results was not warranted, as it was not the primary purpose of this study, the following emerged during follow-up interviews with 15 men and 15 women to elaborate on their survey responses: seven of 15 men and six of 15 women reported having an overall negative body image. For men, their primary concern surrounded the desire to gain both more weight and



LEIGH ANN BRAY, MD

muscle. Primary concerns surrounding body image mentioned by women were 1) happiness with being thin; 2) weight gain having a negative impact on body image; 3) bloating as a concern both pre- and post-lung transplantation; and 4) self-consciousness in public having a negative impact on body image.

To move the initiative forward and gain true insight into these body image concerns, I sought to look deeper and gather information on how processes and practices could be improved to better address these issues. Currently, this study is ongoing, but the preliminary data shared are from individual interviews with seven men and 10 women. The focus of this study was to 1) look into how the clinical variables (such as the use of CFTR modulators, lung function, etc.) were associated with body image and weight; 2) explore knowledge of the impact of weight on overall health status; 3) explore preferences and attitudes of both men and women on concerns and ways to best improve body image; and 4) explore perceptions about BMI/weight measurements as an indica-

tor of health status.

### Body Image and Weight Concerns for Men

During the interviews, most men expressed that they would like to gain both weight and muscle tone. A unique perspective shared included the perception of peers. Some men felt that when they were in positions of authority, in their job or personal roles, that others did not take them seriously. One man mentioned, “I’m a small person for my age, so I just often feel that people don’t take me, my education, my knowledge, or my age seriously, because they think that I’m more in my early 20s than my mid-40s.” Other concerns surrounded lung transplantation, including the emotional impact of nearing lung transplant: “When they brought up lung transplant, that was a great fear, not necessarily scarring, but just to lose an original part that I came with.” After transplant, men expressed the concern of not being comfortable taking their shirt off due to their transplant scar. Overall, men, when facing transplant and post-transplant, expressed a changed view of themselves and found that coping was often difficult.

### Body Image and Weight Concerns for Women

The majority of women voiced that they were content with their weight and did not want to gain weight. However, in some instances, women mentioned the desire to keep their weight as is despite being told by a healthcare provider that they needed to gain weight. One woman offered her perspective: “I feel like in CF, we’re kind of lucky that our body image is more along the lines of what society does than other disease states [sic], obviously.” Other body image and weight concerns were very emotional for women and were mentioned as being associated with stress, anxiety, and/or depression.

These included: 1) peers perceiving them as pregnant due to bloating; 2) peer perception of “looking sick;” and 3) hair, skin, and teeth changes. Some women experienced others misperceiving them as pregnant in their early teens and mentioned that this had a negative impact on their mental health. If they looked sick, both on and off oxygen, and peers and family members commented on their appearance, women said it tended to make them more self-aware and self-conscious. **Subscription Manager**

Lastly, unwelcome changes to hair and skin, together with staining of teeth, involved medications taken to treat CF. Women mentioned that these changes drew heightened awareness of being different from peers and was emotionally very difficult.

### **Impact of CFTR Modulators**

CFTR modulators were mentioned to be “game changers.” Benefits seen included weight gain, stabilization of weight leading up to transplant, improved quality of life, decrease in frequency of

hospitalizations, and an increase in independence. One woman shared the impact that Trikafta had on her life: “I’ve just been healthier all the way around. We call it the miracle drug. I feel like I have nearly no symptoms of CF.” However, an important thought to consider is that not all with CF are currently eligible for existing CFTR modulators. There is still work to be done. Also, the impact that these medications have on body image and weight could change how we, as researchers and providers, can implement changes and interventions now and in the future. We are embarking on uncharted territory in terms of how these medications will impact weight and body image long term.

### **What Does the Future Hold?**

When asked what could be done, the following emerged from interviews: 1) BMI is a good measure but additional information, potentially incorporating muscle mass, could be helpful; 2) providers need to look beyond the numbers; and 3) comments that providers make can both positively and negatively impact body

image. Also, all participants mentioned that body image had either been brought up by their providers “a few times” or “not at all.” Overall, both men and women suggested before or at puberty as the optimal time for the initial conversation about body image to occur with their CF care team.

These insights show that there are areas for improvement and that needs in the era of CFTR modulators are ever changing. It is vital that those who have CF, care providers, and researchers work as a team to move beyond the numbers to formulate and provide the best possible, holistic care to improve weight and body image concerns. ▲

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*Leigh Ann Bray is an Assistant Professor at the University of Alabama at Birmingham. Her research focus is to improve the quality of life of all with CF. Her passion for her research began through her experience as a bedside nurse. In her spare time she enjoys traveling, hiking, and spending time with friends and family. Her e-mail address is: leighannbray@uab.edu.*

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## **NOWAKOWSKI** continued from page 15

need ample vitamins, minerals, and monounsaturated fats from things like nuts or avocados. We also need fluids—not too much, especially for folks like me who have kidney complications, but enough to thrive. If you don’t have CF, this stuff is still important but may have different nuances depending on your circumstances. The bottom line is that doing our best to give our bodies what they need helps us fight off viruses like COVID-19.

**9. Remember that your best is all you can do.** All the items in this list require resources of various kinds. I have a lot of training in sociology and am still very active in that field, so I remain conscious that it can feel easy to say “just do this” if we have privilege of various kinds that makes doing certain things easier. Everyone should do the best they reason-

ably can to practice good infection control and general self-care with the resources available to them. Nothing more, nothing less. If you are doing your best, you are a hero for yourself and many others in these scary times. Trust in that. And maybe think about it for 30 seconds or so while you...

**10. WASH YOUR HANDS!** Soap and water if you can; hand sanitizer or other approved clinical cleansing supplies if you can’t. See the above point about doing your best and knowing your limits. Just wash your hands—whatever they’re made of—early and often, every single day.

Thanks for taking the time to read and reflect on these important ways we can protect ourselves and each other from COVID-19 and other dangerous viruses.

Let me close by saying again that it’s okay to feel afraid. But it’s also absolutely okay to let yourself breathe a little easier. You may not be a superhero in the classic sense, but you absolutely have the power to keep yourself and everyone else a whole lot safer. So use it with confidence! And while you’re at it...wash your hands. ▲

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*Dr. Alexandra “Xan” Nowakowski is a medical sociologist and public health program evaluator. They currently serve as an Assistant Professor in the Geriatrics and Behavioral Sciences and Social Medicine departments at Florida State University College of Medicine. They also founded the Write Where It Hurts project ([www.write-whereithurts.net](http://www.write-whereithurts.net)) on scholarship engaging lessons from lived experience of illness and trauma with their spouse, Dr. J Sumerau.*



# Gaining Weight With Modulators

By Rachel Rone

**I**n 2015, I was enrolled in a trial for Orkambi. At the start of the trial, I was 20 years old, 5'2", and weighed 99 pounds. I had spent my entire life underweight and struggling to gain weight. I have memories of adding Scandishake powder to everything, drinking full glasses of whole milk, eating even after I was full—all in an attempt to get as many calories as possible in a day. The nutritional value of foods didn't take priority. If it had calories, I ate it.

While Orkambi didn't improve my lung function significantly, one side effect that I did not anticipate was the weight gain. Even though I had a hospitalization, I gained 10 pounds the first few months on the drug. The weight gain alone helped resolve a lot of my underlying health issues and allowed my lungs and body to become stronger, even though my PFTs weren't going up as much as anticipated. In the five years since the trial, I have taken Orkambi, Symdeko, and now Trikafta, for the past 4 months, and I have gained almost 40 pounds. This weight gain was great the first few years while in college: I walked from class to class and remained active. However, when I began working full time in July 2017, sitting at a desk most of the day and eating free corporate lunches routinely began to take its toll.

A year into my career, my clothes didn't fit, and I was very unhappy and self-conscious about my body. I didn't want to go on dates or go out with friends because I worried about how I looked. It was a complete shock to feel fat after being severely underweight for 22 years. For my entire life, people had

“ Family and friends made occasional comments about my weight gain, and I knew it was time to make a change. ”

commented how skinny I was and how lucky I was to get to eat whatever I wanted and still remain rail thin. Now I would eat a McDonalds meal and instantly feel bloated and like I had gained 10 pounds. To make matters worse, I had the dreaded CF belly, and I



**RACHEL RONE**

felt that I looked pregnant all the time. Family and friends made occasional comments about my weight gain, and I knew it was time to make a change.

The first step I took was to do research. I scoured the internet for weeks, educating myself on nutrition and healthy eating habits. I also contacted my endocrinologist to get a continuous glucose monitor to understand my blood sugars and learn how certain foods affected my body and sugars. I

decided from the start that I wasn't going to do a crazy diet or restrict myself to only certain foods. I didn't want a quick fix. I wanted a lifestyle change that I could implement and improve on for years to come. I downloaded a food-tracking app to get an idea of how many calories I was consuming, and it was quite the wake-up call. Throughout my research, I learned about portion sizes, counting macrobiotics and additives in prepackaged foods. I couldn't believe how much I didn't know. I had seen so many doctors and nutritionists during my life, yet none of them had taught me these simple concepts. I also called on my friends who were big on healthy eating to get their tips and tricks.

It took some time and self-discipline, but I was able to develop a diet and lifestyle that works well for me. My meals today consist of proteins, vegetables, and healthy carbs with smaller portions of sweet treats. I cook a lot and stick to natural, unprocessed foods whenever possible. I love to use my Instant Pot to meal prep for the week. As an active 25-year-old, I love to go out with friends, so I make sure to look at menus before going to restaurants to see what healthy options I can choose. I'm not perfect, and I still get sugar cravings and eat junk food, but I am more aware now of what I am eating, and I try to make the fast food runs less often.

Once I felt I had my nutrition in a better place, I decided to join a gym

and begin strength training. This was one of the best decisions I ever made. In one year, I have gained so much muscle. So, while I am still gaining weight, it is good weight in the right places! I feel so much better about my body, and I like what I see in the mirror most days now. I have more confidence to go out and meet people and feel good in my own skin, and that is what matters most.

In this new era of modulator

drugs, nutrition will play a big role. Children growing up with CF today have more of a possibility than ever to live long, healthy lives. My hope is that doctors and nutritionists make a conscious effort to teach younger patients how to fuel their bodies with nutritious foods and develop lifelong healthy eating habits. How you nourish your body and a positive mindset can help bring some control during the difficult CF times and lead to bet-

ter health and well-being. ▲

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*Rachel Rone is a 25-year-old living in St. Louis, MO. She was diagnosed with cystic fibrosis (double delta F508) at three months old and CFRD at ten years old. She works full time as an analytical scientist researching new forms of cancer therapies. In her free time, she likes to work out, play volleyball, travel, and attend country music concerts. She can be reached on instagram @rachrone or by email: rachelrone3@gmail.com*

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### **TILLMAN** continued from page 3

occurrence rates using different cutoff values for commonly used laboratory indicators of iron status. Results showed that 41.8% of stable CF patients had iron deficiency, while 33% had anemia. Iron deficiency was more commonly found in women (51%) than in men (30%) with CF. Iron deficiency was significantly associated with anemia, vitamin A deficiency, and antacid use, as well as moderate and severe lung disease, when compared with mild lung disease. In turn, anemia was found to be significantly associated with vitamin A deficiency, severe lung disease, low body weight, diabetes, and bone disease. These factors are generally linked to CF severity, which suggests that anemia may simply be another complication of worsening [CF] disease.

<https://tinyurl.com/rqtp667>

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#### **Chronic Kidney Disease Highly Prevalent In Cystic Fibrosis, Study Says**

Patients with cystic fibrosis (CF) have a high prevalence of chronic kidney disease (CKD) linked to higher levels of uric acid and fats in the blood. These manifestations seem to be particularly severe in patients who receive a lung transplant. As such, performing early and complete screenings in these patients is recommended to determine

the risk of kidney injury, and also of cardiovascular disease. Historically, CF-related kidney disease (CFKD) was considered rare. However, several studies recently have reported different types of kidney problems in CF patients, including in children. Kidney injury can be caused by CF itself (CFTR is necessary for proper kidney function), by other disorders such as diabetes, or by taking high doses of certain medicines, such as aminoglycosides. While acute kidney injury (AKI)—a sudden episode of kidney failure or kidney damage—is well-studied in CF and known to be associated with lung infections and antibiotics, the prevalence and causes of chronic kidney disease (CKD) (a gradual loss of kidney function over time) are more controversial. To shed light on this topic researchers evaluated the incidence and manifestations of kidney disease in 226 CF patients, with an average age of 35 years. The team also searched for possible links between kidney damage and laboratory markers of metabolic disease, inflammation and endothelial dysfunction—dysfunction of the blood vessels that can be associated with a greater cardiovascular risk—and respiratory impairments. Kidney injury was evaluated using the estimated glomeru-

lar filtration rate (eGFR), a marker of kidney function, using the MDRD equation and serum creatinine values. (An eGFR below 90 mL/min/1.73 m<sup>2</sup> indicates kidney damage and was used as a sign of CKD.) Of a total of 226 patients, 65 were found to have CKD. Most of those with kidney damage had a class II CFTR mutation (delF508). Notably, the team found that 28 patients had received a lung transplant, and this was linked with poorer kidney function (lower eGFR) compared to CF patients who had not had a transplant. Lab tests revealed a significant association between worse kidney damage and a greater amount of lipids (fats) in the blood—including a higher level of triglycerides, total cholesterol and low-density lipoproteins (LDL). An increase in serum uric acid (SUA)—a circumstance associated with kidney and cardiovascular events—also was associated with worse kidney function. No relationship was found between chronic kidney disease and blood levels of vitamin D, glucose (sugar) and hemoglobin A1C (a blood measurement of diabetes risk). Regularly testing for eGFR could help predict and monitor kidney damage, especially in transplanted patients.

<https://tinyurl.com/tgzj3gp>

Continued on page 38



## Sunrise: A New Beginning

By Barbara Harison

As I entered 2020, I had made it to another year. Here I am at 73 years old living with cystic fibrosis. I was diagnosed with CF when I was 64 years old! I lost a younger sister to CF in 1971, when she was just 21, but I erroneously assumed I was just a CF carrier who exhibited some symptoms. As I aged, I had more frequent health challenges with bronchitis, bronchiectasis, and pneumonia. Even when I told doctors and pulmonologists about my family history of CF, I was told that I was too old to have it. Finally, I was referred to Dr. Richard Belkin (Santa Barbara Pulmonary Associates), who specialized in adults with CF. After a hospitalization in 2010, with all the genetic testing and other diagnostics done, I was diagnosed with CF. The diagnosis was actually a positive event, all things considered, as I finally have answers for my frequent health challenges and could start treating the underlying cause. I embarked on a whole new regimen of breathing treatments and daily pills—at 64! Thankfully, I am pancreatic sufficient and thus far have not needed enzymes to aid with absorption when I eat. My health improved and I could exercise longer without being short of breath.

I started off January 2020 recovering from both a pulmonary embolism and exacerbation that began back in June of 2019. I was able to get off my blood thinners in December 2019 and weaned myself off the oxygen, but still did not have my normal stamina and lung capacity. For the past five years, Kalydeco has helped stabilize my lung functions, with my FEV1 hovering around 40%. I had very few exacerbations during that time, only one bout of pneumonia, and no more salty skin.



BARBARA HARISON

The 2020 sunrise brought the miracle drug Trikafta and a **New Beginning** of life. It seems too good to be true. I did not experience the purge that many others have mentioned. I had more energy, and, within just a few days, I noticed I wasn't coughing much and I could hike up hills without needing to rest. Within a week, I was back to swimming my laps for 40 minutes at a faster pace without a break. I went to the CF clinic in March where test results confirmed an improvement in my FEV1 and, even better, I did not have any trouble completing the step test. I have experienced some minimal gut side effects and take a daily probiotic to mitigate. I am healthier now than when I was in my fifties.

I look forward to maintaining my active lifestyle and helping others to stay healthy and active. I only wish my sister, Loretta, could still be here to benefit from this new drug. In memory of my sister, I established the Loretta Morris Memorial Fund with the Cystic Fibrosis Lifestyle Foundation (CFLF) 10 years ago. The Fund awards Recreation Grants to CF patients to

help them **Live Stronger and Longer**. Preference is given to grant requests for the activities my sister enjoyed (horseback riding and dance) and my favorite activities (swimming, golf, and yoga). The Fund is supported by donations from 62 family members and friends. Over 10 years \$78,300 has been raised and 170 Recreation Grants awarded to children and adults living with CF.

As I approach the rest of this year I remember that **Everything starts with a Sunrise but it's what we do before it sets that matters** (K. McGraw). I am looking forward to a May trip to Utah to visit the National Parks and hope I can do well at the higher elevations but, just in case, I will take my personal oxygen concentrator (POC) along with me. ▲

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*Barbara M. Harison is 73 years old and has CF. She and her husband, Rich, live in Ventura, CA. She is retired after a long career in public parks and recreation administration and consulting. When she is not swimming laps, beach walking, or golfing she volunteers for community organizations and currently serves on CFLF Board of Directors.*

## Gratitude

I had to do it or I would die.  
I might die anyway,  
but I had to take the chance.

They said "Yes" without even  
being asked.

On May 24, 2000, my uncle Rickey  
and my friend Jerry went into  
surgery,  
and had part of their lung removed.

They were put into me.  
I lived.  
They are fine.

There are no words to describe...  
feelings too deep to express.  
All I can do is say "Thank You."

— *M. Thompson, 2002*



PHOTO BY STEPHEN BOYER

# FROM OUR FAMILY PHOTO ALBUM...



**LARISSA GUILIANO GETTING TO SKI POST-TRANSPLANT AT DEER VALLEY RESORT, UTAH, IN MARCH 2019.**



**MALCOLM CAISE (BROTHER), MAHALIA CAISE (SISTER), MARIAH CAISE, AND FATHER, FIELDING CAISE.**



**LARA GOVENDO DROPPED INTO THE MAGICAL GRAND TETON NATIONAL PARK DURING HER SOLO CROSS-COUNTRY TRIP IN 2018.**



**SONYA OSTENSEN IN COUNTY CLAIRE, IRELAND, DURING A FAMILY VACATION IN 2018.**



**DEVIN WAKEFIELD AT BLUE GLACIER, ON MT. OLYMPUS, WASHINGTON, IN JULY 2019.**



**ANDREA EISENMAN AND STEVE DOWNEY ON VACATION IN JAMAICA IN MARCH, 2020.**



# My Consistent Battle With Weight Gain

By Larissa Giuliano

I have always struggled with gaining weight due to cystic fibrosis. Like many others with CF, I can't digest fats very well and, as my lung function decreases, my metabolism increases. Added to that, I was a competitive athlete as a child and teen and have been a vegetarian (who occasionally eats seafood) since I was eight. I have tried to gain weight in many different ways and have found a few things that work for me.

## Diet

Before I was diagnosed with CF at age two, my dad, who is very athletic and almost 200 lbs., says I could eat as much as him. Despite eating that much, I failed to grow normally. Then I started taking enzymes and, while still thin, I started growing. Around the age of seven, I got the "you really need to gain more weight speech" at every doctor visit. It was frustrating because it's not like I wasn't trying. I tried many different supplements, drank half and half, ate lots of ice cream, and even replaced water with sugary sports drinks (recommended by the dietitian) but could not gain enough weight.

Then I was threatened with a feeding tube if I didn't gain 10 lbs. by my next CF appointment. Horrified by the thought of not being able to swim every day, I was determined and actually able to do it. In addition to high-calorie meals and snacks, I drank several 600-calorie Scandishakes a day, often waking up in the night to chug one down. I have found this to be the easiest way to gain weight without a feeding tube. The shakes are only about 10 oz once mixed with milk and taste fairly good. They are simple to prepare and I can drink one quickly without

feeling too full.

I'm fortunate that my mom is a nutritionist so I grew up eating lots of fruits, veggies, whole grains, and nutritious foods. As I got older and my sister developed CF-related diabetes ("CFRD"), I became more aware of my carb intake. Before then I hadn't really considered how many carbs I ate at a time. I started trying to eat more high-protein, high-fat, low-carb foods and less high-sugary foods. Since I don't eat



LARISSA GIULIANO

meat, dairy has played a large role in my diet. I used to drink somewhere around five cups of milk a day in order to get enough protein. I currently still drink milk but mainly rely on beans, nuts, peanut butter, all kinds of cheese, Greek yogurt, cottage cheese, and eggs for high-fat, high-protein sources. I usually also have protein bars handy to eat on the go or after a workout.

## Feeding Tube

When my health declined significantly throughout college, I lost weight and could not gain it back. I was doing

everything possible to just maintain my weight, including drinking three Scandishakes again. Inching closer and closer to transplant and realizing I needed a certain BMI to be considered, I agreed to get a feeding tube. I had local anesthesia during the placement procedure and spent one night in the hospital before going home the next day. My stomach was sore for about the first week. After a month of the standard feeding tube, I went to the doctor to get it swapped out for the low-profile MICKEY button, which I liked *much* better. The set-up was pretty straightforward. I used the Kangaroo Joey pump and mostly ran it at night while sleeping. I enjoyed not having to wake up in the middle of the night to drink a shake. It also took the pressure off eating. If I didn't feel like eating, which became increasingly frequent the sicker I got, I could use the feeding tube. I felt a sense of relief. With the extra calories at night I gained the weight needed to be put on the transplant list.

It did take some getting used to, though. Sometimes I woke up feeling full and nauseated. At this time, I was on home IV antibiotics, which also contributed to nausea. Adjusting the rate and amount of tube feed helped with this issue. Being pre-diabetic, I needed a low carb feeding tube formula and this proved difficult to find. I found Glucerna 1.5 to be the highest calorie, lowest carb option. My blood sugars rose due to the extra carbs, and I required an intermediate-acting insulin to cover my tube feeds. Maintaining the site was pretty easy—I kept a small split sponge under the button and changed it about twice a day.

While I only had a feeding tube for a year, many times it helped tremendously. A couple months after its place-

ment, my lung collapsed resulting in a drop in my lung functions, down to the 20s. No doubt my weight would've dropped considerably without the feeding tube. During my transplant surgery, my vocal cord was paralyzed; therefore, I could not swallow correctly. In addition to nutrition and liquids, I received all my meds crushed up and through the feeding tube. Although wary at first, I am definitely grateful I had a feeding tube at this time in my life.

### Exercise

My doctors have always advised exercise as part of a daily airway clearance routine. My parents put me in sports from a young age and they have been a huge part of my life, particularly gymnastics and swimming. As I became more and more involved in competitive sports, it became harder to gain weight.

Until age 12, I swam five to six days a week and would often come home from practice exhausted and without an appetite. With all that swimming, I burned more calories than I could eat. When I switched sports from swimming to gymnastics, my appetite seemed better and I built more muscle, allowing me to keep my weight up.

It has been challenging to find the right balance of exercising enough to clear my lungs while not losing weight. I think a combination of both strength and cardio exercise is the best way to be as healthy as possible. When I am not feeling well, I opt for mild exercise like a walk and have learned not to overdo it. Now being post-transplant, I still have to make sure to consume lots of calories when I exercise. However, having an improved appetite makes it much easier

to get in enough calories. My normal routine consists of strength exercises four times a week at the gym where I coach gymnastics. I walk or jog with my dog a couple times per week, swim in the summer, and hike occasionally.

Weight gain has been an undesired adversary in my CF battle but there are so many supplements and things that can help. Since 2015, when I received my transplant, it has become easier for me to maintain my weight, despite exercising more and having the same level of malabsorption issues. Thankfully, I no longer burn thousands of calories just to breathe. ▲

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*Larissa is 29 years old and lives in Brentwood, California. Her hobbies include gymnastics, swimming, hiking, baking, and spending time with her dog, Gracie.*

## Jeannine Ricci July 10, 1971– February 1, 2020

**W**e are deeply saddened to announce the passing of Jeannine Ricci on February 1 at 48 years of age. She was an extraordinary director of *CF Roundtable*, joining us in 2017.

Jeannine was an exceptionally kind person who worked tirelessly for the CF community. She was instrumental in enhancing our organization in so many ways. Through her dedication and by increasing awareness regarding clinical trials and mental health issues in our community, among other initiatives, she helped numerous adults with CF receive the latest therapies and treatments. She provided hope for a better future and put many of us on the road to better health.

It was always enlightening to work with Jeannine.



She was very insightful and presented unique perspectives while discussing different aspects of CF. She was very engaging, thoughtful, articulate and precise in all she did for us—writing, fundraising, clinical trials and website surveillance.

Her loss is profound, yet her legacy will continue in our programs at *CF Roundtable*. We feel privileged to have worked side by side with Jeannine and will miss her greatly. A beautiful handwritten memorial letter from her

loved ones, Mr. and Mrs. Hurst, states: “If Jeannine is not with the Good Lord, then none of us has a chance; she was that good. May she rest in peace.” Please keep her family, friends and many loved ones in your heart and prayers.



## Life As A Vegan: Over 100 Days Eating A Strict Vegan Diet And I'm Not Looking Back

By Anonymous

I was diagnosed with CF at six months; I had a bilateral lung transplant in 2013; I had my first kidney transplant in 2018; and a second kidney transplant in 2019. Despite these challenges, I have consistently been an advocate for my own health, doing my own research, planning, and investigating all things related to CF. *CF Roundtable* has been a significant resource in this endeavor. Self-advocacy requires strict compliance in my daily medical regimen, both pre- and post-transplant. The only reason I am alive is because I followed my intuition in 2009 that lung transplant was my only hope for a better future. Had I not planned ahead, researched transplant hospitals and doctor choices, and completed my evaluation long before transplant, I would certainly be dead.

I was one of those patients who fell *fast* into the state of lung failure; so fast that I was on the transplant waiting list for only eight days. In the fall of 2012, I had low, but stable, lung function, performing all my normal life activities, albeit with some difficulty. My FEV1 was in the 30s (with oxygen at night only). I was going to Pittsburgh for transplant follow-ups once a year to check my lung status. I was due to go to Pittsburgh on December 13 for a yearly transplant check-in. At this point, I was not yet on the transplant list. As I was eating Thanksgiving dinner that year with family, I started having acute pain in my chest. Less than a month later, I was on life support and ECMO and was given just a few days to live. Because I had been proactive, I had already completed my lung transplant evaluation before this sudden collapse in lung health occurred. My determination to stay in

control and be the advocate for my own health helped in part to save my life. I say “in part” because there are many other factors that contributed to my lung health: my faith in God, support from my husband through all of my transplants, prayers, and an astounding medical team. Did I mention my husband donated one of his own kidneys to me only six months ago?

The above story of how I fought to

“ About two weeks after my diet change, I started noticing that my stomachaches were less frequent and less severe. ”

recover my health serves as an introduction to the story of how I embarked on a vegan diet. This story is pertinent because I like to ask questions and seek answers as much as my limited medical knowledge allows. When I first considered veganism, I read as much research as I could get my hands on in addition to talking with my care team and transplant doctors. All of this research led me to one conclusion: I wanted to try to be vegan again.

I had tried being vegan once in 2005 with little success. I was then living alone in an expensive city (Los Angeles) without any family support. The amount of effort it took to prepare vegan meals, and the cost of alternative products, was almost impossible and I did not succeed. What I couldn't foresee, however, was that, by 2020, the market for vegan products would change significantly and there would no longer be just one substitute for each animal product. And, even better, prices for these alternative products

would be trending downward as new products were created due to the increase in demand.

Today there are numerous different milks and non-dairy creamers, not to mention the huge variety of prepackaged vegan burger options. And, they're tasty! Access to homemade recipes for *any* entrée you want to make can be found on the thousands of vegan websites, apps, and cookbooks that provide non-animal

alternatives to pretty much anything you want. Even our local specialty hamburger bar in my tiny town outside Buffalo has a fabulous vegan option!

I decided to try veganism again in December 2019, because I kept reading more and more articles about choosing plant-based foods to potentially reduce your chances of getting cancer, which now, as a transplant patient, is my main worry. Rejection is always a concern after transplant; however, I have lived for 7.5 years with new lungs and have yet to experience any rejection, so I see cancer as being the main reason to change to a strict vegan diet. For me, eating vegan means: no animal meat, poultry, cheese, milk (or any dairy), eggs, or fish. The only exception I make is honey.

I knew the change was not going to be easy but, after being reminded of the benefits of plant-based eating from watching recent documentaries on Netflix, I knew the time was now or never. My husband was on board as well after educating himself and committed to the

dietary overhaul the same day I did.

Although I knew I might not *feel* the difference in the change in diet right away, I was shocked to experience significantly less arthritis in my hands after only three weeks. Arthritic discomfort had been a constant irritant during the 12 months prior to starting my vegan diet. After 100 days of being vegan, I have virtually no more arthritis pain in my hands.

Most importantly, I cannot believe the difference it has made in how my stomach feels. For the last three decades, I felt like my stomach was constantly in turmoil, as if I was constantly on the verge of a sour stomach despite taking all my enzymes and vitamins as prescribed. I always took my enzymes at the same time (at the beginning of a meal) and, as an adult, never forgot them. I could never pinpoint either the specific food causing my stomachaches or the determining factor(s) in their severity. Many times, I had such bad stomachaches that I could do nothing after eating a meal except either bend over, or sit down, until the pain subsided. About two weeks after my diet change, I started noticing that my stomachaches were less frequent and less severe. In fact, I started needing fewer enzymes with each meal. For every pre-vegan meal, I needed to take five Creon enzymes. Now, I eat two or three with breakfast and four with dinner. I have not had any stomachaches since changing my diet—none.

Prior to being vegan, going to the

bathroom was another issue that constantly caught me off guard. Not only was I on a completely sporadic schedule no matter how many regular meals I ate, but my bowel movements were never the right form and consistency. As personal as this discussion is, I had no idea how much my gut was out of whack until I completely changed the foods I was eating.

Weight was another question. I did not know if I would be able to sustain my weight. After eight weeks of my vegan diet, I had not lost one pound. In fact, it remained exactly the same, even with my consistent exercise routine of five days per week, which included 25 minutes of cardio, yoga, and weights! I will be honest, though, and admit that, post-transplant, I did not have trouble staying at a healthy weight, either. I was still concerned, though, that I might lose weight because of the change. Surprisingly, I have stayed at 119 pounds since changing my diet and continuing to exercise.

I will note that the one item I did have an issue with is gas and/or yeast infections, because I no longer ate yogurt. Since I am taking two antibiotics prophylactically post-transplant, I need to replenish the good bacteria in my gut. Vegans who are eating a lot of raw fruits and veggies, combined with a high-legume diet, can be stricken with bloating and gas. The solution I discovered for myself is a juice full of probiotics from my regular grocery store.

I really can't say for sure if it was the removal of dairy, or perhaps eating a much more balanced diet full of fruits and veggies, or maybe a combination of all the dietary changes that has contributed to my calmer stomach! Whatever the true scientific explanation is, I am never going back to eating animal products for as long as I can help it.

To be honest, I will miss getting my custard cones this summer, but I am committed and willing to keep eating my dairy-free Dole Whip instead, which I love just as much. I have never been tempted to "cheat" because my husband and I are doing this challenge together. I do think it would be much more challenging, if not impossible, if my husband was not on board with this new diet.

I started this mission to help reduce my chances of getting cancer. Whether this will ultimately help, I will truly never know. I do know that it has helped reduce my inflammation, my stomachaches, and unpredictable bathroom trips. The cherry on the top of this adventure is that my food choices are not only kinder to the planet, but they also help save animals. I am more than willing to make the change because, so far, the benefits far outweigh the sacrifice. ▲

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*You may contact the author with any questions about this article by e-mailing [cfroundtable@usacfa.org](mailto:cfroundtable@usacfa.org). Please reference this article in your e-mail. All responses will be answered by the author and returned anonymously at her request.*

## Cystic Fibrosis Mothers

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

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

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



# Controlling My Weight

By Andrea Eisenman

For those of us with CF, weight can be such a touchy subject. Because I have been rail thin most of my life, with the ability to chow down on large amounts of food numerous times a day, all without gaining weight, it was hard for me to get a grips on my current situation, which requires me to control my weight sensibly.

I am not overweight, but I'm definitely heavier than I would like. I am not sure if my ideal body image is skewed from weighing less than 100 pounds most of my life, but I do have a muffin top that I hate, and I feel flabby at times. Despite exercising to stay in shape, both physically and mentally, there are certain areas that remain stubbornly intact, like my tummy, face, and butt. Like others with CF, when I was trying to keep weight on in my 20s and 30s, I had to eat several times a day and snack constantly. For a while, I enjoyed all the unlimited food. Eventually, I found this task difficult: it only took one or two hefty coughs after overeating before it was a mad dash to the nearest bathroom or waste basket to sadly throw it all up.

My constant eating at work seemed to frustrate my co-workers, who were struggling to lose weight. They hated that I had dried fruit and nuts plus weight-gain shakes at my desk, in addition to consuming massive amounts of food at lunch. Those who knew my struggle tried to understand but, still, the thought bubble over their head intimated, "she should be grateful she is that thin without trying." I later realized that weight can be an issue in anyone's life as it is one of those things over which we can have some control, or perhaps it gives us a sense of control when we are content with our weight and appearance. In the end, what else can we control in life? We

cannot control our genes or how they are expressed, obviously, nor what family we are born into, etc. But we *can* have an effect on our weight and body image, which can make us feel good about ourselves—like an achievement that we worked hard toward.

After I went on disability, and was listed for transplant, I tried almost anything I could think of to gain weight,



ANDREA EISENMAN

to little avail. I didn't lose weight, but I was teetering at around 90–93 pounds. I became frustrated with my lack of progress to gain, and I was assessed for a feeding tube for night feedings. Both times I was about to have the surgery to implant the g-tube, I ran a fever and the surgery was canceled. After that, my CF nurse told me they could train me to put a tube down my throat to use for night feedings, and then I would not need the surgery. I was desperate and went a few times to the doctor's office to insert a thin tube down my nose into my gut, which would attach to a machine to tube feed me at night. As you can imagine, inserting the tube made me gag and cough uncontrolla-

bly. Finally, I told them I would just try to drink high-protein drinks and avoid snaking a tube down my esophagus. It helped me to maintain but there was no gaining. And with CF, during exacerbations, it is better to have a few extra pounds for just such occasions. But because I was in a constant state of exacerbation—respiratory failure—any weight gain was fleeting.

Fast forward to post-transplant life, and I was still able to eat everything I wanted and, more importantly, I no longer had coughing fits after a full meal, and thus I was spared any immediate purging afterwards. I was beyond thrilled and appreciated my new ability to eat the foods I enjoyed without repercussions. I happily accepted this new development. Then I felt my metabolism change when I hit 40. I had maintained my weight between 100–105 pounds after transplant but, after my metabolism slowed, I started to feel like I was putting on weight rapidly. And I could not stop my eating habits that had been ingrained in me my whole life: eat, eat, eat, and eat some more. And my appetite was fierce.

Speeding ahead again to my late 40s, I crested at about 135 pounds. I hated how I looked. Because I am short, I looked, in my opinion, a bit like a meatball. My transplant doctor told me: better to be heavier so if you do get an infection, you will be able to fight it off more easily and not waste away. I was not wasting anything, especially not food! I want to attribute some of my healthy weight gain to being diagnosed with diabetes right after transplant and starting insulin injections. Since I was likely pre-diabetic—if not fully diabetic—prior to transplant, my body wasn't fully processing any food I ate, and all my efforts to gain weight were fruitless

if my sugars were high. Adding to that, as I entered perimenopause in my 40s, my metabolism came to a grinding halt and I was forced to restrict my eating and watch what I ate—gulp! I was mortified and could finally see the other side of the coin—trying to lose weight. I didn't like it. Again, I was reminded how out of control I felt when all I wanted to do was eat despite being uncomfortable with the extra weight and feeling restricted by my clothes at times. I even went to an Overeaters Anonymous (OA) support group at a friend's urging. I eventually realized that food driving my appetite wasn't the sole cause of my weight gain—an antidepressant called Remeron (generic name Mirtazipine) was a contributing factor as well. When I was on that for depression and hyponatremia (a low concentration of sodium in my blood), my appetite was tenfold and out of control. In the past 20 years, I have been on this medication on and

off, but mostly on as it helps me sleep. Do I want a good night's sleep or do I want to be slim again?!

Once I realized this, I remained on Remeron with a renewed goal: try to stop grazing constantly, typically post-dinner. I tried to remain conscious of whether I was eating due to anxiety or actual hunger. I learned from my brief research and OA meetings that sometimes we eat our feelings. When we are sad, anxious, angry, etc., we can take comfort in self-soothing. For me self-soothing looked like food, but for others it looks like alcohol, drugs, sex, and so on—pick your poison. So, after eating a full bag of potato chips or having three servings of dessert after dinner, I became angry with myself and the self-loathing ensued. After each binge, I vowed to start anew the next day, knowing I had to change my behavior. It took a lot of time with a lot of failure along the way. Eventually, persistence in making good food choices put me

on a better path for success.

I am now through menopause and no longer feel as out of control with my weight or eating. Sure, I do go on eating jags, but I no longer beat myself up about it. I have learned, like most humans, to eat slower and stop when I start to feel full. The learning curve was steep—I started to see that breaking old patterns that had served me most of life—eating a lot and often—were not going to benefit me or my health as my body changed into my later years. I do feel more in control of my weight and body now. This allows me to feel less anxious, thereby stopping the need for self-soothing while also rejoicing in my small accomplishments in maintaining my weight. ▲

*Andrea is 55 and has CF. She lives in New York, NY, with her husband Steve and dog, Willie. She enjoys cooking new recipes, playing pickle ball, biking, tennis when possible and staying active as her health allows. Her contact is a Eisenman@usacfa.org.*



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# IN THE SPOTLIGHT

## With Mariah Caise

By Andrea Eisenman and Jeanie Hanley

**M**ariah was hospitalized, fighting a lung infection, when I, Andrea, first interviewed her, and I could tell immediately just how determined she was to fulfill her life's goals. These include teaching, finishing her Master's Degree in Accounting, and possibly becoming a forensic accountant at the FBI! She doesn't let her disease keep her down but is realistic and works around it to live a full life. She doesn't want to be seen as sick or use her disease as an excuse. Each day, Mariah makes a conscious effort to ensure she consumes the proper amount of calories thanks to a recent g-tube placement, even while at work. Her motivation will impress you, so read on to learn all about our newest star. Spotlight please!

**Age:** 23

**Where do you live?**

I live in East Brunswick, New Jersey. I have lived here my entire life, and I live with my father, sister, brother, brother-in-law, and our pet dog, Max.

**How is your health?**

My health has been a rollercoaster. My baseline FEV1 is usually between 65%-75%. But I have been hospitalized and on intravenous antibiotics off and on this past year. I have an antibiotic-resistant pseudomonas that makes it tough to treat, but even more tough for me to stay healthy once I'm off antibiotics. The pseudomonas has become resistant to almost all antibiotics except for one, Tobramycin. The downside with Tobramycin is the loss of hearing as a side effect—I am now completely deaf in my left ear and my right ear has 35% hearing loss.

I have also recently experienced tachycardia, which is an elevated heart rate. It feels like my heart is racing even when I'm just sitting down. For now, it is not of major concern to my doctor



MARIAH CAISE

because I have had infections and inflammation, which is most likely the cause of it, but it is being monitored and I now see a cardiologist. I was also diagnosed with gastroparesis, which is slowed or delayed emptying of my digested food. This has caused weight issues and pancreatic issues such as constipation and bowel obstructions. I have trouble gaining weight, so I had a feeding tube put in a little over a year ago. This helped reduce the stress of having to consume large amounts of calories every day when I feel full.

I have also been hospitalized quite a few times; in the beginning, I would get PICC lines to administer my medications, but soon my veins became too scarred or would clot from the PICC lines, so I had a port placed. This was a great decision because it's one needle poke and it's done. The needle has to be changed weekly but it is more reliable, less painful, and more convenient as it can be used to administer drugs and take blood. This is a huge bonus because I have small and wiggly veins

so when a peripheral blood draw is required, it usually takes two to three times to hit the vein.

**Tell me about trying to get a bachelor's degree and what you did when you had to drop out?**

My schooling situation was unexpected. I graduated high school in 2015, and I was excited to go to the University of Kentucky. Here is what I thought I would do for four years: work towards a nursing degree, eventually working in pediatrics or oncology for the rest of my life. Well, things did not go according to plan. While away in Kentucky for my freshman year, I was hospitalized six times, and even though I still managed to have a 3.6 GPA for the year, I decided that Kentucky was not the right place for me. The doctor I had in Kentucky actually said that my lungs were not adjusting to the air down south as they were accustomed to the New Jersey air.

So, I transferred to Rutgers University in New Jersey and commuted from my house—this only lasted one semester. The struggle of having to walk to class every day, wait at the bus stops, and just commuting generally made it difficult to fit in treatments because I'd be on campus all day. Also, during the winter season, it was difficult to stay out in the cold for long periods of time while walking to class or waiting at the bus stop.

As I transitioned to Rutgers, I switched majors from nursing to pharmacy. I had a reality check and realized that being a nurse and the exposure to sick patients all the time would affect my health. However, I wanted to stay in the medical field so I chose pharmacy for my new major. After one semester I once again got sick, missed almost two months of class, and had to withdraw

for the semester. After this, I took a semester off, worked full time as a manager at Taco Bell, and thought about what my next steps were.

I eventually decided to attend school online because this would give me the flexibility to complete schoolwork from anywhere (at home or in the hospital) and work to earn money without worrying about class schedules interfering with my work schedule. My two favorite subjects were always science and math. Science didn't work out for me, so I decided to choose a career in the math field and accounting was the perfect choice. This was the best decision I made. I completed my undergraduate degree online at Excelsior College in two years because I took classes year-round with no breaks. After graduating, I decided to earn my master's degree online as well. I applied to the University of Michigan-Flint and was accepted—I was elated, and now I am currently working on my Master's Degree in Accounting.

### **What are your goals for the future once you get your MA?**

While completing my master's program, I applied for the FBI internship program, which occurs in the summer. I have not learned the results regarding the internship but when I complete my master's program, I plan on taking the CPA (certified public accounting) exam. The exam requires 150 credit hours, which the master's degree will fulfill. I would like to work for the FBI as a forensic accountant, but I have other possibilities. My secondary option would be to become a tax accountant, and while I am engaged in these jobs, I would like to return to school online to obtain my doctorate degree because, in the long-term future, I would like to teach accounting at a university.

### **How will being an accountant affect your schedule? Will you work from home?**

Working as an accountant would

be beneficial for me because I can work remotely or at a desk job in an office setting. I don't have to stand on my feet all day and I can do treatments while working. Also, many accountants have varied work hours. For instance, oftentimes tax accountants work a lot during tax season but then their hours wane in the off-peak season. This would afford me the time to focus on my health—it ensures a balance of work and health.

### **How has work helped or hindered your aspirations?**

Currently, I work as a manager at Taco Bell. The job has offered me accommodations in the form of flexible hours, working certain shifts while avoiding others, like the night shift, so I can manage my health. They have also given me breaks so I can do bolus tube feedings during the shift and essentially keep my energy high. And the job is flexible—when I go into the hospital for exacerbations, I have a job to come back to without worrying about being let go. Not only did they pay 50% of my undergraduate tuition, they also gave me a scholarship for \$3,000 during my undergraduate study. This helped me study without being in debt, which was a blessing.

### **When you were diagnosed with depression, what helped?**

During my time at the University of Kentucky, I was diagnosed with depression. Mental health is just as important as physical health. Numerous hospitalizations while at the University of Kentucky took a toll on me mentally. I was sleeping a lot, not going out with friends, missing class, and overall not happy. While in the hospital I was irritable and refused to talk to the doctors. It was then a doctor diagnosed me with depression. The depression hit a low when I decided to take the semester off from Rutgers and then withdrew. I thought I was a failure in life and felt like I was disappointing every-

one around me, mainly my family.

I decided to start going to therapy sessions with my social worker. These sessions helped me set goals and get to the root of my unhappiness. It brought up unresolved feelings about my illness and my mother. My mother died from cancer in 2014, so she never got to see me graduate, go to prom, celebrate my eighteenth birthday, etc. I never really coped with that and I think this caused prolonged anger and confusion. The therapy sessions were helpful and made me see that I have a purpose; that I cannot let myself go down this path of giving up. I took charge of my life, made goals, made a plan, and stuck to it.

### **What do you do for fun? And in your free time?**

In my free time, when I'm not working, doing treatments, or doing schoolwork, I like to watch TV. My favorite shows to watch are *Criminal Minds*, *Grey's Anatomy*, and reality television. I also like to crochet. I have crocheted scarves, mittens, headwraps, and even blankets. It's a great way to pass the time and the items turn out nicely. I also like to do Sudoku puzzles because they challenge my mind while also being fun to complete. I also like to play cards, such as UNO, Solitaire, War—count me in for any card game. I am not a homebody; I like to go out with my friends. I love to go bowling, golfing (mini-golf or top-golf), play games at Dave & Buster's, and walk the beach and boardwalk during the summer.

### **What motivates you to keep pushing to succeed?**

My main motivation to keep pushing are my parents. My mother was diagnosed with cancer in 1998 and she fought that terrible disease for 16 years until the cancer took over her body. She is my motivation to never give up and, more importantly, not let the disease dictate my life. My father has always been a caretaker for either my

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## CF: THE MIND GAME

# When Lightning Strikes More Than Once

By Mark Tremblay

I was born in San Jose, California, in 1969; however, when I was six, I moved to Apalachin, a small town in the Susquehanna Valley. Like most small towns in upstate New York, it has its share of both hard-luck stories and local heroes that have made the valley proud. My mom has made the town her home for more than four decades. She is beloved throughout the valley for her superior seamstress skills and, as such, loves to share all the town gossip with me whenever I come home. Because my mom has a penchant for telling great stories, I always look forward to hearing them. One of these stories was about a softball pitcher in the valley who played for the varsity team as a freshman in high school. Her ability to direct her 60-mph fastball into the top inside corner of the strike zone at will caught the eye of scouts from nearby colleges. According to local lore, she was struck by lightning, not once but twice. The first time—and this was more than 35 years ago when she was a freshman—she was struck on a hot and humid July day while playing with friends. The jolt knocked her unconscious, so she was rushed to the hospital, where she was revived in the emergency room. She was left with chronic joint pain, loss of her natural athleticism, and an inability to recover her groove on the mound. In her junior year, she was struck again, but this time she was cured, her athleticism was restored and, most notably, she recovered her groove on the mound: she could hurl a 65-mph fastball, which earned her a full scholarship to one of the best teams in the country.

Although I never could verify the story, it deeply resonated with me as a

teenager because I took huge risks. I rationalized that, as a CFer, I had already drawn the shortest of short straws in life's lottery and therefore nothing worse could or would happen to me—essentially, lightning never strikes more than once. This flawed notion spurred many of the stupid decisions I made as a teenager: jumping out of the bed of a pickup doing 80 mph onto an open trailer in the adjacent lane; picking a fight with the assistant captain of the college hockey team while visiting on a college tour; and, most notably accepting a friend's dare to see if his three wheeler could crest the highest wall of the quarry down the street. For much of my life that dearly held delusion that lightning never strikes more than once proved mostly true as I enjoyed a certain level of success despite my poor health and increasing CF complications.

When I turned 38 all that changed—I was caught in a metaphorical violent storm that struck me with one bolt after another, forcing me to abandon my long-held delusion about the reoc-

currence of lightning. Within a 15-month period, I lost the job I loved in the state capital, my 16-year-old marriage ended, and I was routinely hospitalized for CF exacerbations and complications. To make matters worse, the mortgage crisis of 2008 led to the bank foreclosing on all of my properties, including my home, forcing me to declare bankruptcy.

As you might expect, I sunk into an all-consuming depression that was so overwhelming, I could gather neither the energy nor the desire to do anything about it. I felt like I was going through life half-asleep and without my faith, family, and friends, I don't know how I would have made it. Previously, I had always made things happen; but I was quickly becoming a victim of life's happenstance. Luckily, I managed to find a job and an apartment, so I could keep trudging on, despite the fact that I had definitely lost my groove.

Life became a lot quieter and lonelier in the following months, which afforded ample time to emotionally convalesce and reflect on the role I had played in making a mess of my life. I started individual counseling with my former marriage counselor, changed my anti-depression medication, and started listening to country music as it now spoke to me in ways that it never had before. My counselor quickly surmised that, in addition to my serious underlying health condition, I also struggled with low self-esteem. According to her, I masked my feelings of self-worth by an insatiable need to prove myself, and the resultant emotional tension had contributed greatly to many of my failed relationships. One day, deep into an early-morning session, my counselor asked me if I believed that, "...I am fearfully and wonderfully made" (Psalm



MARK TREMBLAY

139:14). I answered by not answering. In reply, she said something that turned the tide of my depression: “If you’re going to be the hero of your own story, like that softball pitcher your mom told you about, you’re going to have to confront your most destructive, deep-seated core beliefs that living with CF has ingrained in you.”

She was right! In my 38 years of living with CF, the disease had not only wreaked havoc on my body, but it had also infected my mind to the extent that my mind had become the single biggest obstacle I had to face if I was ever going to find my groove again. Below are a few of the toxic core beliefs I had to overcome if I was going to be the hero of my own story:

- I didn’t deserve to live because so many people far more worthy than I had long since passed;
- I was a cost rather than a benefit

to society because of how expensive my medical care is;

- I was such an emotional drain to people that I did not deserve to be loved by others;
- I was a blight to the gene pool that needed to be eradicated to better the human species;
- I needed to constantly strive to be better than everybody else in order to prove my worth; and
- I was too sick for anyone to want to marry me for better or worse.

I realized that those toxic core beliefs were no less delusional than my prior notion that lightning never strikes twice. In fact, those toxic beliefs had become proverbial mental termites, eating their way deep into my psyche—I could no longer mentally withstand the hard storms of life. I realized that I had to identify and exterminate all of my CF-related mental pestilence if I was

ever going to recover from this setback.

Before the next storm hits, I encourage you to take a few minutes to reflect on whether you’ve got any destructive core beliefs that may make it more difficult to not only withstand the next storm but outlive it. Ultimately, you get to decide whether the next storm is the catalyst that could return you to the mound as a hero. ▲

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*Mark is 50 years old and lives in Albany, NY, with his wife MaryGrace and stepson. He holds an M.A. in Psychology from Marywood University and an M.P.A. from Syracuse University. Mark has worked for six years in the New York Governor’s Division of Budget and currently works full time at the Department of Health. He and his wife love cycling, church ministry, and riding their motorcycles. You can follow them on their YouTube channel, “Breathing Grace.”*

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**CAISE** continued from page 33

mother or me. He helps me in all aspects of my life, including setting up my medications for me, organizing my feeds, and helping with treatments by either setting up the nebulizer or clapping on my back for reinforcement after the vest treatment.

I want to be healthy and adhere to my treatment regimen, not only for myself, but for my father. I want to live a long life so I can provide the same care to my father as he did for me. I never use cystic fibrosis as an excuse to not be able to do something; I want to show others that they can do anything they set their mind to, despite having an illness. I also don’t want people to pity me or treat me differently because of my illness. CF is an illness I have, but it doesn’t have me and, more importantly, it doesn’t define me. I may have to move a little slower, take a longer path

to get to my goal, or modify certain activities, but I will still achieve my goal at the end. CF will not stop me; it may try but I am the boss of this disease.

### **How has the feeding tube helped you?**

Getting a feeding tube was a great decision for me. I have had trouble gaining weight my whole life, and, once it’s gained, it is hard to maintain the weight. I used to have anxiety about seeing my weight when going into clinic because even losing two pounds is a huge issue. With a feeding tube, I am able to give myself calories during overnight feeds and boluses during the day to add more calories into my diet. The thought of having to stuff my face and consume the recommended caloric intake (3,000 calories daily) was a weight lifted off my shoulders.

### **Do you have a motto you live by?**

My life motto is “If God brought you

to it, He will bring you through it.” If there is one thing that has gotten me through life and dealing with this illness, it is my faith. Not everyone believes in God, but I do. I believe God looks out for me and gives me the strength to fight this illness and allows me to be an inspiration and role model to others, showing them that everything will be okay. There will be pain and hardships along the way, but it is how you get through it that allows you to come out stronger. ▲

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*Jeanie Hanley is 57 and is a physician who has CF. She is a Director and the Treasurer of USACFA. Andrea Eisenman is 55 and has CF. She is a Director of USACFA and is both the Webmaster and Executive Editor of CF Roundtable. Their contact information is on page 2. If you would like to be interviewed for “In The Spotlight,” please contact either Andrea or Jeanie.*



## LIVE OUT LOUD

NEW  
COLUMN

# Sharing Your Story: The Power Of Vulnerability

By Lara Govendo

When pondering a name for my column, I thought about the goal for my message to the world and it became clear as day: *live out loud*. Since I began the journey of openly talking about my life, the nagging voice inside my head keeps pushing me to share my story. Is it fun? Being vulnerable rarely is. Is it important? Yes, because, in doing so, it tells others they are not alone. I believe our stories don't belong to us—they are meant to be shared so we can connect on a deeper level and find the common ground that unites us as humans.

Living out loud became important to me when I first began telling others that I have cystic fibrosis. Growing up, nobody knew. My parents chose to keep it confidential from the school system to prevent bullying or labels being put on me. I'm beyond grateful for their choice—I was already bullied enough for being too skinny. Telling others that I had CF would not have been well received with the plastic faces of inauthentic people at school.

I didn't want to wait until people saw me with oxygen tubing lacing my face around the clock, so before beginning the evaluation for a double lung transplant, I started talking about my life with CF: the good, the bad, and the ugly. From that point on, my life goal has been to be open about the life I lead; to be the same person on the inside that I am on the outside, in every environment. I work hard at this every day. It's definitely uncomfortable, but the rewards outweigh the discomfort. Living my truth is liberating and I wouldn't want to live any other way.

The growing pains from allowing

everyone into your sphere of living are real. The gut punch of fear that kicks in just before publishing a piece I've written about my life hurts on various levels. Risking judgment, misperceptions, and hate mail are valid. Nothing hurts worse, though, than feeling like you are all alone in your individual daily battles. Baring your soul for the world to see and connect with is worth the cost of vulnerability. It affords us the opportunity to share the grief, gifts, and celebrations that are brought forth by humanity on the deepest level. And I will never regret or apologize for that.

We live in a time where everybody shares the sunshine, butterflies, and happy moments of their lives on social media, while keeping the blood, guts,

and gore behind closed doors. It's rare to get a snapshot of the authentic side of people's lives. Trying to one up each other, the comparison trap, and air-brushed portraits prevent us from connecting with one another in a real way. It's time to bring authenticity, relatability, and connection back again. It all starts with you and me....

I invite you to join the journey and *live out loud* with me. Whatever that looks like for you, I challenge you to dig deep within yourself and find the real you. The person you are inside (before everyone around you told you who to be). And then share that with the world. I believe there's a message we can learn from each other's individual lives and purposes. We need more people who are willing to take the risk and be unapologetically themselves; to share messages of hope, connection, and vulnerability with everyone. I hope that you will step out and be the difference, bring light to dark situations, and resonate with those you encounter on a deeper level—as human connection was intended! ▲

*“Baring your soul for the world to see and connect with is worth the cost of vulnerability.”*



LARA GOVENDO

Lara Govendo is 33 years old and has CF. She lives in Vermont as a wild, adventure enthusiast who holds a Master's Degree in Mental Health Counseling. She writes about living out loud and develops educational programs to restore hope to those in need. Thanks to her double lung transplant in 2017, you can now find Lara traveling on the regular, exploring the glorious outdoors, and belly laughing with her loves. You can find her online at: [www.laragovendo.com](http://www.laragovendo.com) and on Facebook and Instagram at "Lungs4Lovey". Her email is [lgovendo@usacfa.org](mailto:lgovendo@usacfa.org).



# SAVORING SERENDIPITY

## A Gratitude Journal

By David Tarnow

In these seemingly tumultuous times, it can be difficult to find and keep our collective center. There is a lot of noise out there on all media outlets about the coronavirus. I've gotten several messages in recent weeks from friends and loved ones simply dropping a line, inquiring about how I am doing. While it is undoubtedly a very serious situation and we, as a population of high-risk patients, need to exercise an abundance of caution, I do hope that we can all find ways to foster and nourish a sense of peace and gratitude in our daily living.

Personally, I have a lot to be thankful for. Life looks very different for me today than it did 18 months ago. For context, in addition to the daily treatments, frequent tune-ups, and hospitalizations many of us CFers endure during the course of our adult lives, I was home bound, requiring oxygen 24/7, having to do infusions every 12 hours on an indefinite basis, and not physically able to do much of anything on my own. Ordinary acts like taking a shower put me out. I had lost quite a bit of weight and eating was more or less a non-starter. One hour of pulmonary rehab with no perceived benefit and an insufferable amount of effort led to the need to sleep for about four hours after each session.

Consequently, I did not have a lot of hope this time last year. My body was weak and ailing, my spirits were low, and I certainly harbored a lot of fear and anger. My mind had reverted to "survival mode," and I know that I was not very fun to be around. Having

to reckon with the prospect of dying at a relatively young age is not easy and I did not think it fair. Revisiting such dark places in my memory is not enjoyable, but I imagine it to be a beneficial exercise in the study of contrasts and one that will lend perspective to the present moment.

However, a light came over the horizon about six months ago when I was able to start taking Trikafta via compassionate use. Fast forward to today and life is good in a manner of speaking. I still need to use oxygen at

night and with activity, but not all of the time. My appetite has reappeared, and I am now able to enjoy eating again. The medication burden is no less than it was before Trikafta, but I have been given the gift of stability. All of these are victories in my book.

It's funny how tides can turn and inevitably do so. Such is the way. I am learning to appreciate the contrasts. Just as the natural seasons change, so do the seasons of our lives. As we enter into spring, I hope to be more like the flowers and trees of the fields—embracing

the rain when it falls and learning to grow from it. Likewise, when the days are long and the sun is shining, I hope to remember to look up and find my center in the light.

This newfound life has been nothing short of a miracle for me and

has allowed me to alter my trajectory and mindset in ways I desire to maintain going forward. If there is one major takeaway from my experiences over the last couple of years, it is that life goes on. Questions regarding the meaning of life continue to dominate my psyche, and I'm still not so certain that I can answer them with confidence. However, with this new lease on life, I am simply choosing to live with hope, gratitude, and patience. This, to me, is what it means to savor serendipity. ▲

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*Questions regarding the meaning of life continue to dominate my psyche, and I'm still not so certain that I can answer them with confidence.*

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DAVID TARNOW

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David is 31 and has CF. He lives in Erie, Colorado, and enjoys playing the acoustic guitar, learning new skills, deep conversation, good coffee, and getting to the heart of matters. He can be reached at [dtarnow@usacfa.org](mailto:dtarnow@usacfa.org).

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# Meet A New Director — Lara Govendo

**M**y name is Lara and I am 33 years old with CF. I am a native of upstate New York and currently reside in Vermont. I received my Bachelor of Arts in Philosophy, with a concentration in nonviolence and business administration, from St. Bonaventure University. I received my Master's Degree in Education, specializing in Mental Health and School Counseling, from St. Bonaventure University as well. I have always been fascinated with people's personal stories, believing there's always a reason someone is the way that he or she is—you just have to dig a little deeper to find out!

Writing has always been a therapeutic outlet for me. In recent years, I have started openly sharing my journey with CF, transplant, and this road called life. You can also find my column, "Valiant Voice," on the Cystic Fibrosis News Today website, in addition to my regular

blogs for the Cystic Fibrosis Lifestyle Foundation.

I was relatively healthy until my late 20s, when my health quickly tanked in the spring of 2017. By August of that year, I had undergone a bilateral lung

transplant. Forever grateful for my new lease on life, I adopted the motto "Live Out Loud," which also happens to be the title of my new column in *CF Roundtable*. I am honored to be a director and excited to connect with new people, learn from others. Connecting with other CFers and transplant recipients has proven to be a meaningful way to find camaraderie.

I'm an avid adventurer and nature is my happy place. I enjoy kayaking, mountain biking, and hiking, to name a few. I am blessed with a wonderful support squad of family and friends who appreciate belly laughs, tasty food, and adventuring on the regular, like I do. My background in mental health lends to an appreciation of deep conversations and a passion for bringing hope to those who are struggling. You can connect with me on Facebook and Instagram @Lungs4Lovey or on my website: [www.laragovendo.com](http://www.laragovendo.com). ▲



LARA GOVENDO

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**TILLMAN** continued from page 21

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## Thoracic Spine Osteoarticular Pathology With Lung Involvement Common In Cystic Fibrosis

Patients with cystic fibrosis show significantly higher rates of thoracic spine osteoarticular pathology than the general population, with notable correlations between the osteoarticular irregularities and lung involvement, according to a study. Both clinicians and radiologists should be aware of the higher risk for thoracic vertebral osteoarticular pathology in this patient population. Thoracic spine should be systematically assessed in chest radiographs and CT scans to detect early changes in order to implement adequate conservative measures to prevent a more advanced involvement. The study results showed a significantly higher prevalence of pathology compared

with rates of the general population described in the literature. The findings underscore that "there is a significant correlation between the degree of lung involvement and the likelihood of osteoarticular pathology, although their clinical course might not be necessary related. The performance of periodic thoracic CT and bone densitometry studies in cystic fibrosis patients should be considered in order to monitor bone involvement and assess the risk of developing fractures.

<https://tinyurl.com/wrn9w3>

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## Modifier Gene May Explain Why Some With Cystic Fibrosis Are Less Prone To Infection

Researchers have discovered that genetic variations dampening expres-

sion of a gene, called RNF5, offer a likely explanation for why some people with cystic fibrosis don't develop lung infections as early or as frequently as others. In the study the team found that individuals with cystic fibrosis who carry specific genetic variants lowering expression of RNF5 have more mutant CFTR protein on their cell surfaces. Even if the CFTR protein isn't totally functional, it's likely better than having none at all. The RNF5 gene is located in the Major Histocompatibility Complex (MHC). Genes in this region encode molecules that are displayed on the surface of most cells in the body. They play an important role in how the body responds to infections. Scientists have long known that everyone has their own set of MHC gene variations,

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# Meet A New Director — Sonya Ostensen

**M**y name is Sonya Ostensen. I am 45 years old with cystic fibrosis. I was diagnosed at 12 when I had chronic digestion and failure-to-thrive issues. During my senior year of high school, I had an upper-left lobectomy but continued with my education until I graduated from Ohio State University with a bachelor's in environmental science. After working in environmental health for 11 years with the State of Florida, my health deteriorated, and I retired on disability. Feeling the need to be "productive," I volunteered to manage payroll, taxes, human resources, and insurance for my husband's civil engineering firm.

Although my PFTs have declined over the years, I have managed to keep my original lungs. I have participated in many drug studies, including the Vertex drug trials for Symdeko. However, I switched over to the new modulator, Trikafta, after FDA approval in November 2019. The results have been nothing short of miracu-

lous for my overall health, as both my PFTs and energy have increased!

I live in Melbourne Beach, Florida, with my husband of 15 years. We have a wonderful 6-year-old daughter. My life was complete with her seemingly impossible birth and now I have both the best

and most challenging job ever—being a mom. We also love our furry companions—three cats and a pup. My hobbies and activities include gardening, baking, yoga, making jewelry, walking the beach, traveling with my family, climbing trees, and riding bicycles with my daughter.

Having had limited engagement with fellow CF patients in the past, the *CF Roundtable* newsletter has been my connection with others journeying through similar experiences. I am excited to contribute and be a part of this amazing organization, as well as getting to know my fellow colleagues. I started reading *CF Roundtable* more than 10 years ago when my CF coordinator suggested the publication. *CF Roundtable* has been so resourceful, inspiring, and comforting; I have often laughed and/or cried through many articles relating to various writers' experiences. It is inspiring and heartening to know we are not alone on the CF journey—as we Breathe One Day at a Time. ▲



**SONYA OSTENSEN**

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and that they make people more or less susceptible to infections or autoimmune diseases. But because the genes present in the MHC region are so dense with variations, they have not been well-studied for their direct link to diseases. The researchers took a new approach to analyzing MHC gene variants by grouping them. That allowed the team to more easily identify associations between genetic variation, gene expression levels and their effects on complex diseases. The researchers applied this approach with whole genome sequencing of induced pluripotent stem cells derived from 419 people. That translated into investigating more than 4,000 traits, from which they identified 180 associated with variants in the MHC

region. This study uncovered a new aspect of cystic fibrosis—one that could lead to new drug design and development and allow clinicians to better tailor treatments.

<https://tinyurl.com/vkrckpn>

AND

<https://tinyurl.com/vlqs7jq>

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## Presence Of Heme B In The Lungs Linked To Lung Function In CF Patients

The presence of heme B—a component of red blood cells—in the lungs is linked to worse lung function in people with cystic fibrosis (CF). Lung infections with the bacteria *Pseudomonas aeruginosa* are one of the main causes of disease worsening and mortality in

people with CF. *P. aeruginosa* produces chemical compounds called phenazines as a result of its microbial processes. Phenazines are known to be virulence factors—molecules that allow the bacteria to proliferate and spread within the host. The team found a statistically significant correlation between sputum heme B levels and ppFEV1. The data show that the greater the heme B concentration, the lower the ppFEV1 is in CF patients, and a sign of worse lung function. The presence of heme B in sputum samples, according to the researchers, probably results from the presence of blood. Overall, the team concluded that given that hemoptysis is strongly associated with airway inflam-

Continued on page 40

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# Meet A New Director — Devin Wakefield

**H**i, I'm Devin Wakefield, and I'm so excited to join USACFA as a director. I am 29 this April. As a newborn, I was immediately diagnosed with a bowel obstruction (meconium ileus), which led to my CF diagnosis. Such a sudden and intense start to CF proved challenging for all, but learning about CF sooner rather than later ultimately had its advantages. As I grew up, my family and I tried to stay on top of each new obstacle CF threw our way. Despite our best efforts, some issues still hit like a truck on a highway at 2 a.m. Bowel obstructions often necessitated trips to the ER at all hours of the night. As my CF worsened with age, the frequency and severity of multiple symptoms of hemoptysis intensified and,

subsequently, those early morning visits to the ER increased in number. Despite all that CF has thrown at me, I'm still happy I'm alive and able to enjoy this world's delights. Today, I am strong, healthy, and benefiting from the triple modulator, Trikafta.

I love to explore the outdoors, embrace my gay self, and complain about CF. Running helps me explore where I live. I am training for a half marathon and choose routes that help me learn what nearby neighborhoods have to offer. I also love backpacking—the freedom to pick up camp and march wherever I please helps diminish my fears. When I'm backpacking, I have a plan of where to go next; I have power over my destiny. Some of my fears relate

to being gay: growing up gay in a heteronormative world can make one feel especially alone, even more with CF in the picture. The challenges I've faced with CF have had a lasting impact on my perspectives on what matters in life, enabling me to connect more easily with my co-marginalized family and learn from their experiences. Hearing others in the LGBTQ+ community share how they faced abandonment and loneliness gave me a deeper appreciation for my family's enduring, if sometimes flawed, bonds—I never needed to fear getting kicked out, always had support for health care and insurance. To be free, you need support. With support and sturdy ground beneath, you can go where you want, explore every-

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**TILLMAN** continued from page 39

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mation, pulmonary exacerbations, and impaired lung function, these new data suggest that heme B may be a useful biomarker of CF pathophysiology. <https://tinyurl.com/wzjf3w7>

## ***S. Pseudopneumoniae* Bacteria Caused Pulmonary Exacerbations In Cf**

*Streptococcus pseudopneumoniae* should be included in the panel of opportunistic bacteria causing pulmonary exacerbations among those with cystic fibrosis (CF). *S. pseudopneumoniae* is a recently described species of bacteria that belongs to the “mitis group,” within the larger viridans group streptococci (VGS), a varied group of bacteria that colonize several tissues and organs, including the airways. Researchers reviewed clinical and microbiological data associated with *S. pseudopneumoniae* in a group of 20 CF patients. They found that *S. pseudopneumoniae* was associated with pulmonary exacerbation, either as the sole opportunistic pathogen or as part of a polymicrobial

infectious process. Additional analyses showed that *S. pseudopneumoniae* was susceptible to several anti-biotics, including amoxicillin, cefotaxime, pristinamycin, rifampin, vancomycin, and teicoplanin, and resistant or less susceptible to erythromycin, tetracycline, and penicillin. They suggest that these results warrant the need for additional studies to increase knowledge of the epidemiology and clinical significance of *S. pseudopneumoniae* in CF. The team also noted that suitable laboratory tests should be developed and implemented in routine practice to prevent misidentification of bacteria species in patients' samples, and the use of unsuitable antimicrobial treatments.

<https://tinyurl.com/sb5xdlg>

## **Faster CF Diagnosis, Better Treatment May Lie In Noninvasive XV Technology**

A new non-invasive tool, called XV technology, allows professionals to visualize airflow in living lungs and could help in the diagnosis, monitoring, and

treatment of cystic fibrosis (CF).

The technology was developed by researchers at Monash University. Current ways of diagnosing respiratory lung diseases, such as pulmonary function tests and imaging techniques (chest X-ray, CT scans, MRIs), can have drawbacks. While some of these tests are poor at detecting early stages of lung disease, others can require sedation or radiation doses, or lack sensitivity and resolution. Since pulmonary function tests are measured at the mouth, these tests are unable to localize where in the lung any change in function originates. Additionally, CT scans, while providing quality 3D images, cannot image the lung while it is breathing, which means airflow through the airways and into the lung tissue cannot be measured. Airflow and lung function can be measured using phase contrast X-ray imaging (PCXI), an imaging technique shown to provide sensitive and high-resolution images of lung tissue, and one that can track airflow through the

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thing, and find your best self.

Complaining about CF keeps my mental health afloat. Unexpectedly, though, it has also helped me build lasting relationships and connections with others in the CF community as we sometimes struggle to find the right spaces to vent and release frustrations that others struggle to understand. Venting together created some of the most meaningful bonds I've ever created. We realized we faced similar struggles, and that discovery meant we weren't alone. We could learn from each other. We grew stronger knowing we had each other's back. These bonds keep me going with a full spirit in hard times.

I have read *CF Roundtable* for many years now, and I enjoy reading every



**DEVIN WAKEFIELD**

CFer's news and perspectives. Before joining USACFA as a director, I volunteered with USACFA, moderating the Facebook groups and page, posting interesting

CF-related news. In that time, I got to know several of the current directors very well—their exuberant and determined personalities convinced me to join the board as a full director. I think I have a lot to offer through my experiences as an adult with CF, volunteering for various CF organizations, and through my more recent experience getting to know USACFA more deeply. I look forward to keeping the website current, continuing with my role as a moderator on our Facebook page, and writing a few articles and blog posts now and then.

I currently live in Seattle, Washington, and work for Microsoft as a computer engineer. Feel free to say “hi” at my e-mail address: [dwakefield@usacfa.org](mailto:dwakefield@usacfa.org). ▲

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airways. PCXI detailed images allow “the application of four-dimensional X-ray velocimetry (4DxV) to track lung tissue motion [lung expansion and contraction throughout the breath] and provide quantitative information on regional lung function. But PCXI, while a validated imaging technique, requires high-tech synchrotron facilities. (A synchrotron is a particle accelerator that allows the study of different materials and living matter.) According to the team, a 2D version of this technology is currently under review by the U.S. Food and Drug Administration for diagnostic purposes in a clinical setting.

<https://tinyurl.com/vn7djxy>

AND

<https://tinyurl.com/sflq36r>

AND

<https://tinyurl.com/tn8bxvo>

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### **Azithromycin Treatment Outcomes May Differ Based On Antibiotic Type**

Patients with cystic fibrosis saw a greater increase in FEV1 when given

oral azithromycin in combination with certain IV antibiotics for the treatment of *Pseudomonas aeruginosa*, as well as an increased overall improvement of FEV1. The findings, although inconclusive and unable to establish a causal relationship, suggest that concomitant azithromycin may be unhelpful when combined with intravenous tobramycin. In this retrospective cohort study, researchers analyzed the use of azithromycin in combination with IV colistimethate or tobramycin for the treatment of pulmonary exacerbation caused by *P. aeruginosa* in patients with cystic fibrosis. The primary outcome was total lung function recovery as measured by FEV1 following treatment. The analyses, although inconclusive and unable to infer causality, suggest that chronic azithromycin treatment may provide modest additional benefit during intravenous antibiotics to treat a pulmonary exacerbation in those treated with a colistimethate-based drug regimen but not when intravenous tobramycin is

used. Whether or not azithromycin use directly influenced the antimicrobial effects of intravenous colistimethate or intravenous tobramycin cannot be understood from these data. The investigators stated that more research is needed to determine whether azithromycin alters the efficacy of IV antibiotics used in the treatment of *P. aeruginosa*. <https://tinyurl.com/umue57z>

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### **Dangerous Bacteria Communicate To Avoid Antibiotics**

Researchers are trying to develop new types of antibiotics that can fight bacteria, and at the same time trying to make the current treatment with antibiotics more effective. They are now getting closer to this goal with a type of bacteria called *Pseudomonas aeruginosa*. In a new study, researchers found that the bacteria send out warning signals to their conspecifics when attacked by antibiotics or the viruses called bacteriophages which kill bacteria. When

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# Bene factors

## BRONZE

**Anonymous** (in memory of Jeannine Ricci)  
**Anonymous**  
**Irene Bernaix**  
**Ann Black**  
**Brandywine Global** (in memory of Jeannine Ricci)  
**Holly Byrnes**  
**Teri Davis**  
**Lisa & Jeff DeMatte**  
**Amy Ganzel** (in memory of Jeannine Ricci)  
**Cathy Garifalos** (in memory of Jeannine Ricci)  
**Jean Jennings** (in memory of Jeannine Ricci)  
**Emily Kramer-Golinkoff**  
**Haddon Heights Soccer Booster Club** (in memory of Jeannine Ricci)  
**Kathleen Hurst** (in memory of Jeannine Ricci)  
**Anthony Kaleta** (in memory of Jeannine Ricci)  
**KPMG c/o Bergen County United Way** (in memory of Jeannine Ricci)  
**Pat & Tom Lamb** (in memory of Jeannine Ricci)  
**Mansion Avenue School** (in memory of Jeannine Ricci)  
**Ryan McCabe**  
**Cantor Rabinowitz**  
**Michael Ruth** (in memory of Jeannine Ricci)  
**Jessica Sanchez** (in memory of Jeannine Ricci)  
**Sameet Shah** (in memory of Jeannine Ricci)  
**Steven Shin** (in memory of Jeannine Ricci)  
**Betsy Siddon** (in memory of Jeannine Ricci)

**Gail Steele** (in memory of Jeannine Ricci)  
**Jon & Shari Stuckey** (in memory of Jeannine Ricci)  
**Catherine & Dennis Sweeney** (in memory of Jeannine Ricci)  
**David Tattersall**  
**Tinges Family** (in memory of Jeannine Ricci)  
**Francina Webb** (in memory of Jeannine Ricci)  
**Greg Zripko**

## SILVER

**Mark Juelis** (in memory of Jeannine Ricci)  
**Joseph Kirby** (in memory of Jeannine Ricci)  
**Ron & Barbara Kramer** (in honor of Jeanie Hanley)  
**Celia Rodgers**

## GOLD

**The Ryan Girls** (in memory of Jeannine Ricci)  
**Anujeet Sareen** (in memory of Jeannine Ricci)

## PLATINUM

**Foundation Care**  
**Ozmatic, Inc.**  
**Translate Bio**

## SUSTAINING PARTNERS

**Maxor**

## PEARL SUSTAINING PARTNERS

**Boomer Esiason Foundation**  
**John & Susan McMurry Fund** (in memory of Peter McMurry)

**TILLMAN** continued from page 41

they receive the warning signal from their conspecifics, they can be seen in a microscope moving in a neat circle. It is a smart survival mechanism for the bacteria. If it turns out that the bacteria use the same evasive maneuver when infecting humans, it may help explain why some bacterial infections cannot be effectively treated with antibiotics. The next step is to research how to affect the bacteria's communication and warning signals.

<https://tinyurl.com/r428u2d>

### Could Synthetic Molecules Provide A General Treatment For Cystic Fibrosis?

A new treatment for lung disease in cystic fibrosis (CF) could potentially benefit all patients. Researchers aimed to restore transmembrane anion movement by identifying new molecules which transport anions efficaciously across cell membranes. The team designed synthetic "anion carrier" molecules to do the same job as the missing CFTR. Using specialist equipment, the team tested the synthetic molecules'

efficacy in cystic fibrosis cells and found they were not only effective in transporting anions across the cell membranes, but also supplement the effects of new drugs targeting faulty CFTR by allowing more anion transport by cells than either the molecules or new drugs alone. These results suggest a new approach to treat cystic fibrosis. The next stage in this research will be to test the identified molecules on sheets of cells from cystic fibrosis air passageways to determine whether they restore mucus move-

ment and bacterial killing.  
<https://tinyurl.com/vjl15tx>

### Antabio Awarded \$4.4M For Continued Research For CF Lung Infection Therapy

Biopharmaceutical company Antabio has been awarded \$4.4 million by CARB-X to continue its research for a therapy against *Pseudomonas aeruginosa* lung infections in people with cystic fibrosis (CF). The funding will be used to support Antabio's *Pseudomonas* Elastase Inhibitor (PEi) program, which is seeking to develop a small molecule to stop the action of the LasB elastase protein. That protein, made by the bacteria *Pseudomonas aeruginosa*, causes damage and inflammation to lung tissue, and prevents the immune system from defending against the bacteria effectively. Antabio's innovative PEi program seeks to develop an inhaled product to be used as an adjunct to existing therapy and which will aim to reduce the severity of *P. aeruginosa* disease and enhance pathogen clearance by targeting the LasB elastase, a key virulence determinant that contributes to tissue damage and inflammation in infected CF lungs. Antabio believes the PEi product, with its novel target and groundbreaking mechanism of action, has the potential to significantly enhance the effectiveness of existing treatments for CF patients. What makes the Antabio's molecule candidate so unique is that it's not an antibiotic. Antibiotics typically work by killing bacteria, whereas this molecule aims to disable the bacteria, making it less able to attack and in-flame the lungs. By using the molecule alongside the traditional antibiotic treatment, it also makes the bacteria less likely to become resistant to drug treatment. The molecule would be administered alongside the antibiotic treatment in an inhaler form, and patients would likely not even realize their course of treatment has dif-

Continued on page 44

## BOOMER ESIASON FOUNDATION'S PROGRAMS



The Boomer Esiason Foundation Scholarship Program provides assistance to students with cystic fibrosis based on a number of criteria including: academics, athletics, arts, and more...

[www.befscholarships.com](http://www.befscholarships.com)



The Boomer Esiason Foundation Lung Transplant Grant Program provides grants to cover expenses including, but not limited to: temporary housing, food, and transportation costs.

[www.befgrants.com](http://www.befgrants.com)



In light of recent natural disasters, The Boomer Esiason Foundation created a fund that directly assists those affected families in the cystic fibrosis community.

[www.esiason.org](http://www.esiason.org)



Team Boomer is a program that encourages people with cystic fibrosis to incorporate exercise into their everyday lives; provides an avenue for individual athletes in a variety of sports to raise money for cystic fibrosis; and offers assistance to grassroots athletic events looking for a cause to support.

[www.teamboomer.org](http://www.teamboomer.org)



A series of audio and video podcasts, featuring Gunnar Esiason and the Salty Cysters, that highlight people with CF and the challenges they face.

[www.gunnaresiason.com](http://www.gunnaresiason.com)



A series of audio and video podcasts in which Jerry Cahill interviews people with CF who are living, breathing, and succeeding through the power of exercise, nutrition, and compliancy.

[www.jerrycahill.com](http://www.jerrycahill.com)

ferred in any way.

<https://tinyurl.com/vw2xtp7>

AND

<https://tinyurl.com/sjudroh>

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### Industry Collaborates To Screen Existing Drugs To Repurpose For Cystic Fibrosis

Calibr has entered into a two-year agreement with the Cystic Fibrosis Foundation to identify existing medicinal compounds that show efficacy against one of the hardest-to-treat infections for patients with cystic fibrosis. Using its ReFRAME drug repurposing collection—an extensive library of nearly all safe small-molecule drugs shown to be appropriate for direct use in humans—Calibr will focus on treatments for hard-to-treat lung infections. This will include screening for novel antibiotics that may be able to combat any of the 22 bacterial species that comprise the *Burkholderia cepacia* complex. By growing bacteria in a way that imitates the infection environment, and then employing a library of more than 13,000 compounds that are already known to be safe in humans, researchers hope to identify promising antibiotics and accelerate the timeline for developing a drug.

<https://tinyurl.com/vvtdqgc>

AND

<https://tinyurl.com/u60xx4u>

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### Experimental CF Compound ETX001 Shows Promise In Preclinical Studies

Enterprise Therapeutics' experimental compound called ETX001 was able to increase airway fluid secretion and mucus clearance in preclinical models of cystic fibrosis (CF). ETX001 is a potentiator of a chloride channel called TMEM16A. Because TMEM16A's production and function are independent of CFTR—the protein that is deficient in CF patients—the findings validate TMEM16A's activation as a potential therapy for all CF patients, regardless of CFTR mutation. The activation of other channels controlling the amount of salt and water in the airways has been thought to be a potential therapeutic strategy for improving mucus hydration and clearance in all CF patients. TMEM16A, a calcium-dependent chloride channel present in airway cells, is one of these candidates. Researchers validated this hypothesis by boosting TMEM16A's activity through ETX001, a newly discovered and optimized compound.

Exposing airway cells from CF patients to ETX001 increased the cells' fluid secretion, specifically through calcium-dependent chloride TMEM16A

channels. ETX001's effects were independent of inflammation, which was previously proposed to increase TMEM16A activity. Boosting the activation of TMEM16A holds promise as a non-CFTR mediated approach for the treatment of CF, which can be delivered alone or in combination with CFTR-targeted therapies. Enterprise is also evaluating other potential non-CFTR therapies, including those targeting ENaC—a sodium transport channel that is overactivated in the lungs of people with CF—and those focused on cells that produce mucus, with the aim of lessening mucus production and complementing mucus hydration therapies. <https://tinyurl.com/vmtphbn>

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### Translate Bio Receives FDA Fast Track Designation For MRT5005 For The Treatment Of Cystic Fibrosis

Translate Bio, Inc., a clinical-stage messenger RNA (mRNA) therapeutics company developing a new class of potentially transformative medicines to treat diseases caused by protein or gene dysfunction, announced that the U.S. Food and Drug Administration (FDA) has granted Fast Track designation for MRT5005 for the treatment of cystic fibrosis (CF). MRT5005, the first mRNA therapeutic with delivery to the



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**FB CF Connect:** [www.facebook.com/groups/cfconnect](http://www.facebook.com/groups/cfconnect)



**Twitter:** <https://twitter.com/CFRoundtable>

**LinkedIn:** <https://www.linkedin.com/company/us-adult-cf-association-usacfa->

# 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: [cfoundtable@usacfa.org](mailto:cfoundtable@usacfa.org)**

## ANNIVERSARIES

### Transplant

**Zachery Hays, 31**

Vernon, CT

Bilateral lung transplant

3 years on February 3, 2020

lung, is designed to address the underlying cause of CF, regardless of genetic mutation, by delivering mRNA encoding fully functional cystic fibrosis transmembrane conductance regulator (CFTR) protein to cells in the lung through nebulization. The Phase 1/2 clinical trial of MRT5005 is currently ongoing.

<https://tinyurl.com/tu6oel2>

### Crestone Funded By CFF To Develop Antibiotic Against NTM Infections

Crestone has received an award from the Cystic Fibrosis Foundation (CFF) to develop preclinical studies for an antibiotic to treat non-tuberculous mycobacteria (NTM) infections. *Mycobacterium abscessus* (*M. abscessus*) in particular, is an NTM often difficult to treat. Crestone’s program will investigate the use of compounds that can block the activity of MmpL3 (Mycobacterial membrane protein Large 3), a protein that is located on the NTM bacteria’s cell membrane and is essential for the bacteria to build their cell wall. The goal is to demonstrate a pharmacology, tolerability and efficacy profile that warrants the declaration of an IND [investigational

new drug] candidate.

<https://tinyurl.com/v4scm7m>

### Ivacaftor May Reduce Respiratory Infections In Cystic Fibrosis

Among patients with cystic fibrosis, the use of ivacaftor was associated with a 32% reduction in the rate of *Pseudomonas aeruginosa* infection and a 15% reduction in the rate of *Staphylococcus aureus* infection over time. For this retrospective cohort study, researchers analyzed data from the UK Cystic Fibrosis Registry from 2011 to 2016. The database contains information on demographics, clinical care, medication use and health outcomes for patients with cystic fibrosis. In 2016, the number of respiratory cultures and the number positive for *P. aeruginosa* were also available. The primary outcome was the annual prevalence ratios for each cystic fibrosis pathogen and secondary outcomes included time to infection in those who were previously not infected and time to *P. aeruginosa* clearance in those previously infected. Results showed that, among patients treated with ivacaftor, the annual prevalence of *P. aeruginosa* decreased from

48.9% in 2013 to 35.9% in 2016, with reductions seen each year. The annual prevalence of *S. aureus* also declined from 2013 to 2016, although the decreases were less pronounced (33.3% in 2013 to 30.1% in 2016). The prevalence for *Aspergillus* spp. also declined 12% to 4.7%, but the change in prevalence of *Burkholderia cepacia* was less than 1%. Although the study is a retrospective study, it shows promise for ivacaftor to improve clinical outcomes for patients with cystic fibrosis. If these drugs are taken before chronic infection starts, the risk of developing infection in the future may be reduced considerably. Also noted was the fact that the study adds to the evidence supporting the long-term benefits of ivacaftor.

<https://tinyurl.com/thgnves>

### Kalydeco Improves Lung Function In CF With Non-Responsive Mutation, Case Study Reports

Treatment with Kalydeco (ivacaftor) alone improved lung function and quality of life in a cystic fibrosis patient carrying both a therapy-responsive and a non-responsive mutation to treatment. The patient, a 35-year-old woman, carried two CF-causing mutations: the Q493X, a class 1 mutation that is not responsive to therapy, and 3272-26A > G, a class 5 mutation that does respond to treatment. This patient had reasonable lung function, but with significant upper lobe bronchiectasis, which is a permanent enlargement of the airways with build-up of mucus accumulation. She was taking a nebulized mixture to treat her bronchiectasis. To help with sputum release, the patient also performed daily airway clearance, and did aerobic exercise twice a week. Treatment with Kalydeco was started as a trial for two months, with clinicians monitoring several parameters at the start and at the end of treatment. According to the Sino-Nasal Outcome Test (SNOT-22) and

Continued on page 46

Revised Cystic Fibrosis Quality of Life Questionnaire (CFQ-R), the patient experienced improvements on both upper and lower respiratory tract health upon treatment with Kalydeco. The woman also experienced improvements in quality of life, especially on physical state, vitality, and health domains of the CFQ-R score. Periodic CT scans showed that the patient had a reduction in mucus accumulation in the lungs after two months of treatment. Lung function also improved after the 2-month treatment period and was sustained for one year following treatment initiation. Further, cardiopulmonary exercise tests demonstrated that the patient improved her workload and maximal oxygen consumption (VO<sub>2</sub>). The data from this case report suggest that the improvements seen with Kalydeco as a single agent on the EXPAND trial could be due to the therapy's effects on the 3272-26A > G mutation—which is responsive to Kalydeco treatment—since the second mutation does not respond to this medication. The team suggested that similar clinical benefits may also be observed in patients with other residual function mutations despite not having an F508del mutation on their second allele.  
<https://tinyurl.com/w8zzhdk>

### Orkambi Therapy Reduces Hepatic Steatosis In CF Patients, Study Shows

People with cystic fibrosis (CF) who were treated with Orkambi (lumacaftor/ivacaftor) have significantly less accumulation of fat in the liver—a condition known as hepatic steatosis—which is often associated with CF. The most common form of CF-related liver disease (CFLD) is called hepatic steatosis—an accumulation of fat in the liver, which can lead to liver damage. It occurs in up to 60% of CF patients, and can progress to cirrhosis, a condition which typically requires a liver transplant as treatment. CFRD occurs in around 20% of adolescents with CF, and in up to 50% of adults. It is charac-

terized by low amounts of insulin, or insulin insufficiency. Orkambi, a CFTR modulator, is a combination of two compounds: lumacaftor, which increases the number of chloride channels, and ivacaftor—marketed as Kalydeco—which helps defective channels work better. No studies have assessed the impact of these therapies on CFLDs such as hepatic steatosis. A research team investigated the prevalence of hepatic steatosis among CF patients receiving Orkambi and those who did not. The team also investigated if there was an association between CFRD and hepatic steatosis. The amount of fat in the liver was measured by magnetic resonance imaging (MRI) to determine the proton density fat fraction (PDFF). A PDFF value greater than 5% was defined as clinical hepatic steatosis. The results showed that the range of PDFF varied between 0% and 21% in the group analyzed. Six of the individuals (30%) had hepatic steatosis based on their PDFF value. All nine patients who received Orkambi had a significantly lower PDFF, ranging between 0.0% and 6.4%, with a middle value of 2.0%, compared with those who had not received the CFTR modulator. Regarding the potential link between CFRD and hepatic steatosis, the results showed that patients with CFRD had a lower PDFF compared with those with normal glucose tolerance. However, the difference was not statistically significant, indicating there was no link between CFRD and hepatic steatosis.

This study raises the possibility that CFTR modulator therapy may impact other forms of CFLD and adds to the small but growing literature on the extrapulmonary impact of CFTR modulator therapy.  
<https://tinyurl.com/wtheepr>

### Organoid-Based Personalized Medicine Trial Enrollment Complete

The Human Individualized Therapy of CF (HIT-CF) initiative will test PTI

drug combinations through the company's CHOICES clinical trial, which will be the first to gauge the potential of personalized therapies for CF. The study will use tissue samples from 502 adults with CF to evaluate three of PTI's cystic fibrosis transmembrane conductance regulator (CFTR) modulators. These modulators act directly on the CFTR protein, which is faulty in CF, to restore its normal function. Tested compounds will include the CFTR potentiator dirocaftor (PTI-808), the CFTR corrector posenaftor (PTI-801), and the CFTR amplifier nesolicaftor (PTI-428). CHOICES builds on early findings from the HIT-CF initiative showing the feasibility of testing these CFTR modulators in organoids. Organoids—in effect, mini organs—are made up of cells grown in a dish. Unlike conventional cells, organoids are grown in specific three-dimensional structures that more closely resemble how cells are arranged within organs in the body. In the first phase of the study, researchers will use biopsies to develop organoids for each patient. All three CFTR modulators will be tested in these organoids to assess efficacy. The results of the tests will be used to define which treatment each participant will receive in the trial itself. During the trial, participants will be given either an active treatment or a placebo, and then switch. The crossover study design allows for greater statistical power, because each participant serves as his or her own control.  
<https://tinyurl.com/vroht6o>

### A CRISPR Cure For Cystic Fibrosis?

Scientists say they're perfecting a safer approach to CRISPR that doesn't involve cutting DNA—and they have early evidence that it could be used to correct a gene mutation that causes cystic fibrosis. The technique, called base editing, allowed the researchers to correct a mutation in the gene CFTR that causes the buildup of mucus in the lungs and other organs in cystic fibrosis.

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But instead of employing the enzyme Cas9 to cut the mutation out of the gene and then replacing it with a corrected piece—as would be done with traditional CRISPR editing—the team used base editing to repair the mutation on-site in stem cells from cystic fibrosis patients. With the new base-editing

technique the mutation in the CFTR gene can be detected and repaired without creating further damage in the genome. The researchers warned that using base editing in the treatment of cystic fibrosis could ultimately prove challenging, because the disease affects more than just the lungs. So far,

CRISPR has shown the most promise in diseases that affect just one organ or tissue type.

<https://tinyurl.com/v5s63t8> ▲

*Laura is 72 and has CF. She is a former director and President of USACFA. She and her husband, Lew, live in Northville, MI.*

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- We would like to act as a referral source for active adult support groups. Please send us your group name, leader's name and phone number, number and age range of your members and geographical area covered, and we will add you to our referral list.
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- Share your ideas for **Focus Topics**, feature articles or any suggestions for improvements you may have to help make *CF Roundtable* more relevant and interesting to you.
- You can reach **USACFA** and *CF Roundtable* at any time by e-mail at [cfroundtable@usacfa.org](mailto:cfroundtable@usacfa.org)
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## IMPORTANT RESOURCES

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

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

**Transplant Recipients International Organization, Inc. (TRIO):** Phone: 1-800-TRIO-386 <http://www.trioweb.org/index.shtml>

An independent, nonprofit, international organization committed to improving the quality of life of transplant recipients and their families and the families of organ and tissue donors. For information, write to: TRIO, 7055 Heritage Hunt Dr, #307, Gainesville, VA 20155 or e-mail them at: [info@trioweb.org](mailto:info@trioweb.org)

**American Organ Transplant Association (AOTA):** Phone: 1-832-930-AOTA (2682) <http://www.aotaonline.org/>  
Helps defray out-of-pocket travel expenses for transplant recipients. Helps to set up trust funds. For more information, write to: Administrative Service Center, American Organ Transplant Association, P. O. Box 418, Stilwell, KS 66085. Preferred method of contact is e-mail: [aotaonline@gmail.com](mailto:aotaonline@gmail.com)

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