

CF CONNECTION

Newsletter of the Vermont Cystic Fibrosis Center Advisory Board

FINDING CALM AMID THE CHAOS



IN THIS ISSUE

Finding Calm Amid the Chaos [pg. 1-2]

CFTR Modulator Update [pg. 3]

The TRIFAKTA Experience [pg. 4]

New Faces in Clinic [pg. 5]

Is Marijuana Safe [pg. 6]

CF Scholarships & Financial Aid [pg. 7]

Coronavirus Information [pg. 8]

by Ashley Mitchell-Ringuette, CCLS

The healthcare setting can be a scary and overwhelming place where stress and anxiety tend to hit a peak. The anticipation around an upcoming clinic visit or scheduled hospital admission can bring up a lot of emotions and questions for both patients and caregivers.

- How are my PFTs? Are they showing a decline in lung function? Will I go home on antibiotics again because of an infection?
- Do I need blood drawn or my port accessed? The smell of rubbing alcohol is such a trigger for me. Oh no, we forgot the numbing cream!
- I wonder how my child, sibling or partner is coping. How can I support them when I am feeling overwhelmed myself?

Relaxation and coping techniques are important tools that help create a sense of calm. Child life specialists, social workers, and mental health experts are vital members of your health care team who can be called upon to help navigate overwhelming feelings. Focusing on relaxation is not about being lazy. Rather, it helps to focus on increasing one's well-being and the mind-body connection.

Below you will find a range of relaxation exercises that can be incorporated into your daily routine or accessed during a clinic visit or hospital admission. They can also be utilized by caregivers: *(continued on page 2)*

VERMONT CYSTIC FIBROSIS CENTER

PEDIATRIC PROGRAM
802-847-8600

Tom Lahiri, MD, Pediatric CF Program Director
 Kelly Cowan, MD, Associate CF Program Director
 Jillian Sullivan, MD, Associate CF Program Director
 Lauren Elizabeth (L.E.) Faricy, MD, Pediatric Pulmonologist
 Keith Robinson, MD, Pediatric Pulmonologist
 Tara McCuin, PhD, Psychologist
 Martine Antell, PharmD, Pharmacist
 Emily DiSchino, RN, CF Nurse Coordinator
 Melissa Barron, RN, Pulmonary Nurse Coordinator
 Rylee Kilbride, RN, Pulmonary Nurse
 Michael Bissonette, RT
 Maryann Ludlow, RD, CDE
 Christine Prior, LICSW
 Maggie Holt, PT
 Ashley Mitchell-Ringuette, CCLS
 Julie Sweet, BA, CF Research Coordinator
 Chloe Housenger, Clinical Research Coordinator
 Lauren Haskins, Medical Assistant
 Kendra Therrien, Practice Support Specialist

ADULT PROGRAM
802-847-1158

Charlotte Teneback, MD, Adult CF Program Director
 Juan Pablo Perdomo Rodriguez, MD, Endocrinologist
 Zachary Weintraub, MD, Pulmonologist
 Tara McCuin, PhD, Psychologist
 Martine Antell, PharmD, Pharmacist
 Abe Sender, PA-C
 Connie Lotspeich, RN, CF Nurse Coordinator
 Erick Duprey, RN
 Kitty Brady, RT
 Maryann Ludlow, RD, CDE
 Sasha Morey, PT
 Christine Prior, LICSW
 Julie Sweet, BA, CF Research Coordinator
 Chloe Housenger, Clinical Research Coordinator
 Lisa Benoure, Medical Assistant
 James Crane, Medical Assistant
 Brent Michale, Medical Assistant
 Jenna Carroll, Practice Support Specialist
 Susan Heney, Operations Support Specialist

Finding Calm Amid the Chaos (continued)

Guided imagery, often called guided meditation, is a relaxation technique that focuses on imagination and use of the senses. Soothing sound or music may be applied alongside a script. Our child life specialists use guided imagery during procedures as a form of distraction. Guided imagery can be used in any setting. Check out apps *Sitting Still* and *Calm* for more ideas on how to use guided imagery. Both apps can easily be downloaded to your phone or tablet.

Reiki is a gentle form of energy healing. Practitioner's hands are placed just over or lightly touching the body. The goal is to promote healing, decrease symptoms, and increase relaxation. A handful of our child life specialists are Reiki II certified, including child life in the Children's Specialty Center and on Baird 5. Ask a member of your team if Reiki is something that you may be looking to try out.

Calming jars are a therapeutic activity and a quick and easy project that can be made at home. There are a variety of ways to create calming jars. Personalization is always encouraged. Swirling around the contents (glitter, beads, etc.) of the jar and watching it settle to the bottom lets children focus on breathing and re-centering themselves, resulting in a sense of calm. Directions for creating your calming jars can be found on Google or Pinterest.

Stress and anxiety can provoke a constant need to fidget. When you are able to focus your attention towards a task or movement, this can help improve a sense of calm and concentration. **Therapy Putty** is a wonderful tool that can be easily transported to and from appointments. Patients or caregivers can manipulate the putty in their hands while engaging in conversations or during an exam with the provider. *Crazy Aaron's Thinking Putty* is a favorite that comes in different textures and colors.

Yoga is known for its ability to help reduce stress and promote relaxation. It can help improve mental and physical health outcomes and can easily be modified based on the person participating. Check out a local studio near you or access free yoga videos online.

CFTR Modulator Update

by Thomas Lahiri, MD

It's here!

I'm sure that most of you are aware of the exciting news: the recent FDA approval of the triple combination modulator, elexacaftor-tezacaftor-ivacaftor (brand name TRIKAFTA™)! With the availability of this drug combination to people with CF who have at least one copy of the F508del mutation, most people will eventually have access to highly effective modulator therapy. Right now, TRIKAFTA is only available to those who are at least 12 years old. We expect safety studies to allow access to younger children in the near future.

What can we expect?

The clinical trial showed some amazing effects of this triple drug combination. Most patients in the study had an average increase in lung function of 13% (FEV1). There was a decrease of pulmonary exacerbations (flare ups needing antibiotics and often hospitalization) by almost two-thirds. Just as importantly, people on this therapy report a significant improvement of their respiratory symptoms soon after beginning treatment. These include looser sputum and less coughing. We know that this drug is able to correct defective CFTR because sweat tests performed on CF patients taking TRIKAFTA are normal or close to normal in most cases. Ultimately, we hope that these effects will prevent the development of lung disease or prevent things from getting worse. It should also help thin the mucus in the sinuses and intestines, which can lead to fewer symptoms and complications.

What don't we know?

There are studies that will be looking at the long-term effect of triple combination therapy. So far, people seem to tolerate this drug very well. It is important to continue to monitor for liver injury with blood tests every three months for the first year, then yearly. Children (less than 18 years old) are required to have an annual eye exam. At this time, we don't know if people will be able to stop some of their other CF therapies. There is a trial beginning soon that will look at this possibility.

The future

There are now four approved CFTR modulator therapies that will eventually treat about 93% of people with CF. This is fantastic, but it leaves 7% of the CF population without any options for modulator treatment. The CF Foundation is working hard with pharmaceutical companies to find an effective treatment for the 7% of people with CF who have Class 1 mutations (also called nonsense or missense mutations). There are new techniques, called gene editing, which may allow for correction of all types of CF mutations, but this isn't available yet. It is also important to note that the goal would be to eventually treat a person with CF right after they are diagnosed, to prevent many of the disease complications from ever occurring.



Patient Perspective

The TRIKAFTA™ Experience

By Sidney Bewlay

I am a 20-year-old patient with CF and I am currently a junior at UVM. I have been on TRIKAFTA for about three months and I have noticed some big changes in my quality of life! Before taking TRIKAFTA, I was on Symdeko for around two years. While Symdeko was helpful, I didn't have nearly as rapid improvement as I do on TRIKAFTA. When I began TRIKAFTA, the first five days felt like a purge from my lungs. I was coughing up an incredible amount of mucus, more than I ever had before. However, it felt similar to when I'm on IV antibiotics and doing my "sick" treatment plan rather than just coughing up mucus because I am sick. After five days, I felt the clearest that I have ever been. As I tell anyone who asks, for the first time in many years I feel like my treatments are working.

TRIKAFTA isn't a cure for me, as I'm sure any patient/parent/care giver knows, but it has transformed my life. This is the first spring for me in the past eight years where I've made it from the fall until now without needing a PICC line. Thanks to this medication (and all of my hard work with treatments/medication regimen), I can laugh again without having a coughing fit immediately after. This semester, I've been able to go to the gym multiple times a week and work out with friends for up to two hours while keeping up with their pace. I have stopped coughing up blood when I work out too and I physically feel much more energetic every day than I have over the past four years. I knew that TRIKAFTA was really changing my body when I went skiing with my family and I was able to pull my dad on his snowboard across a flat without having to take a break and catch my breath!

I'm really excited to see how my health improves over this year. Recently, I had a checkup with the pulmonology team here at UVM and my PFTs had improved from a 67% predicted FEV1 in December to a 75% predicted FEV1 all on their own. My lung function has not raised without assistance from antibiotics or a PICC line in years! It is SO exciting to see correlated improvement to this medication, as well as proof that all of the time and effort I spend on my treatments helps me. Being able to take TRIKAFTA has drastically changed how I live my life. I feel less anxiety over my physical health, I'm more confident about being in school, and I feel more capable in taking care of myself/living on my own as I can see how my health can improve again.

New Faces in Clinic



Maryann Ludlow, RD, CDE

Nutritionist

Maryann has joined the pediatric CF team, replacing Carlie Geer, RD, who retired after many devoted years working with our pediatric CF patients. Maryann has worked in the field of nutrition for over thirty years and has had a rich variety of work experience in the New York City metro area and in Vermont. She attended NYU and CUNY Lehman College for undergraduate studies and CUNY Queens College for an MS in Nutrition Education. She has been a Certified Diabetes Educator since 1997 and has worked with the adult CF population at the University of Vermont Medical Center since 2015. She lives in East Montpelier with her wife and two cats. She loves veggie gardening, kayaking, and cross country skiing, as well as cooking yummy, nutritious dishes.



Chloe Housenger

Clinical Research Coordinator Vermont Lung Center

Chloe joined The Vermont Lung Center in March of 2019 and started working with the CF team in July. She was born and raised in Vermont, but spent some time living and working in Arlington, Virginia while completing her B.A. in Interior Design from Marymount University.

When she's not busy working as a Clinical Research Coordinator or as a waitress at the Upper Deck Pub, Chloe enjoys spending time with her husband Alex, their dog Stout, and their two cats Woodhouse and Kingsley!

CF PROGRAM RECEIVES QUALITY CARE AWARD

The Pediatric Cystic Fibrosis Program at **UVM Children's Hospital** received a Quality Care Award from the Cystic Fibrosis Foundation (CFF). The award recognizes CFF-accredited care centers that have continuously demonstrated a commitment to improving the quality of care they provide to people with cystic fibrosis.

Thomas Lahiri, M.D., Professor of Pediatrics, is Chief of Pediatric Pulmonology. The Pediatric Cystic fibrosis Program just received full re-accreditation from CFF for their site visit.

Is Marijuana Safe for a Person with Cystic Fibrosis?

By Kitty Brady, RT and Charlotte Teneback, MD

There are many ways to “take” marijuana. You can smoke it in a pipe or “joint,” vape it, eat it or swallow it (in a pill form). There is a lot of talk about marijuana - legal or illegal, recreational, medical, grow your own, buy off the street, buy in a “head shop” or order on line. As providers, we are often asked if one form of marijuana is safer than another.

Instead of answering this question immediately, we would first like to review the different forms of using marijuana and the health effects of each.

Marijuana is a plant which, when grown, can attract different bacteria from the dirt. These bacteria include *Pseudomonas Aeruginosa* and *Aspergillus*. Whether smoking marijuana in a pipe or in a joint (rolled cigarette form), you will inhale any of the bacteria the plant may be carrying. Added to this, depending upon where the marijuana came from, you inhale many of the same toxins, irritants and carcinogens into your lungs as in smoking a tobacco cigarette. A carcinogen is a cancer-causing agent. Marijuana is usually inhaled very deeply and held in the lungs longer than a cigarette, causing more exposure to these toxins and irritants. Smoking marijuana can cause a cough and inflammation in the lung airways, which will increase mucus production, making you more prone to lung infections and increased shortness of breath. It can even cause severe types of infection, such as lung abscesses with *Aspergillus*. Per the American Thoracic Society, “smoking marijuana can cause serious harm in people with pre-existing lung conditions.” Second-hand marijuana smoke exposure can cause many of these same issues.

Vaporizers, e-cigarettes, electronic cigarettes or disposable (Puff Bar) are all forms of vaping tools. When vaping marijuana and/or tobacco, you will also inhale as many of the same toxins as smoking a pipe or a joint. In fact, the lung tissue damage found in the lungs of people who died from vaping-induced lung damage was an injury pattern that resembled a chemical burn or a toxic chemical exposure injury. In the past year, there has been a large increase in the number of people who present to hospitals with respiratory failure due to vaping. This has even been given a new name: VALI (Vaping-induced lung injury). Most people who became ill from vaping used a marijuana product.

Vaping also delivers much higher amounts of tetrahydrocannabinol (THC). The higher levels of THC can cause psychotic episodes, hallucinations, paranoia, panic attacks and impaired motor ability.

Edibles come in many forms; gummies, brownies, chocolate... These are not regulated, so the amount of THC in an edible can vary with each batch, depending upon who made it and how it was made. So you really don't know how much THC you are ingesting. It could be a very small amount or a very large amount. When smoking THC, the effects are felt pretty quickly and they are gone in a few hours. However with edibles, it takes one to three hours before you start to feel the effects because they are absorbed through the digestive system first. They peak (feel the strongest effect) in three to four hours. The effect of edible marijuana lasts for many hours. One worry is, if you eat an edible and don't feel the effects yet, you may take some more which makes it easy to overdose on it. There has been an increase in emergency room visits from people who are overdosing from too much THC. An overdose can look like anxiety, panic attacks, rapid breathing, increased heart rate (30 - 50 beats more than normal per minute), low blood pressure, intense paranoia and hallucinations. Sadly, children who accidentally ingest an edible can experience respiratory failure and coma.

There are pill forms of medical marijuana used for nausea and pain, which are prescribed. These are more purified forms of the chemical in the marijuana plant.

These are the facts to consider. You will make your own decision, but please think very carefully first before using any forms of marijuana.



CF Scholarships and Financial Aid

Many scholarships and financial aid options are available for students with cystic fibrosis who want to pursue higher education.

There are many types of scholarships available based on different criteria. For example, scholarships might be based on being involved in athletics, being in a military family, having a skill or ability, having cystic fibrosis, or having a chronic disease in general. When you start your search, it might be helpful to think about different types of scholarships for which you might be eligible.

A good place to start looking for scholarships specifically for people with CF is [Cystic Fibrosis Research Inc.](#), which maintains a list of about 20 scholarships and their eligibility criteria. [NeedyMeds](#) also maintains a list of CF-specific scholarships.

The [Federal Student Aid Information Center](#) at the U.S. Department of Education provides information about finding grants and scholarships, including tips on where to look for them.

The [National Association of Student Financial Aid Administrators](#) has a database of financial aid options that you can search for by state.

In addition to these state and national scholarships, UVM Medical Center is happy to offer **The Vermont Cystic Fibrosis Scholarship**. This \$500 scholarship is provided by a Vermont family. Applications are due by May 1, 2020. To apply, email Christine Prior, LICSW a letter stating your interest at Christine.prior@uvmhealth.org. Recipient's name will be chosen from qualifying candidates at random. Eligible patients are those students who have not received this scholarship previously and are enrolled in a college program for the fall.

If you would like help finding scholarships, call Cystic Fibrosis Foundation *Compass* at **844-COMPASS** (844-266-7277) Monday through Friday, 9 a.m. until 7 p.m. ET, or email compass@cff.org.



The University of Vermont Medical Center
The University of Vermont Children's Hospital
111 Colchester Avenue
Burlington, VT 05401

CF CONNECTION NEWSLETTER

The CF Connection Newsletter is produced twice yearly by the Vermont Cystic Fibrosis Center Advisory Board.

NEWSLETTER EDITOR
Liz Hammel

NEWSLETTER PRODUCTION
Susan Hayward

PEDIATRIC PROGRAM
Children's Specialty Center
East Pavilion, Level 4
111 Colchester Avenue
Burlington, VT 05401

PHONE: (802) 847-8600
FAX: (802) 847-5805

ADULT PROGRAM
Gartman Center for Medicine
East Pavilion, Level 5
111 Colchester Avenue
Burlington, VT 05401

PHONE: (802) 847-1158
FAX: (802) 847-7244

Online Resources

Coronavirus Information

Many of you have reached out to us looking for guidance around how to best protect yourselves from exposure to Coronavirus (COVID-19). Websites from the CF Foundation, CDC and VT Department of Health are below and contain the most up to date information and current recommendations:

CF Foundation: <https://www.cff.org/Life-With-CF/Daily-Life/Germs-and-Staying-Healthy/What-Are-Germs/COVID-19-Community-Questions-and-Answers/>

CDC: https://www.cdc.gov/coronavirus/2019-nCoV/summary.html#anchor_1580064337377

VT Department of Health: <https://www.healthvermont.gov/response/infectious-disease/2019-novel-coronavirus>