FALL 2021

THE University of Vermont MEDICAL CENTER

THE University of Vermont Children's Hospital

CF CONNECTION

Newsletter of the Vermont Cystic Fibrosis Center Advisory Board

COVID VACCINATION AND THE CF COMMUNITY

IN THIS ISSUE

By LE Faricy, MD

The Cystic Fibrosis Foundation (CFF) and the UVM Cystic Fibrosis Center strongly support vaccination for all people with CF and their family members who are eligible for the vaccine.

As of September 17, the CFF reported vaccination rates from the CF Registry that were much lower than expected:

- 35% of eligible patients over the age of 16
- 20% of eligible 12-16 year old patients

While we would like to assume these numbers are from under-reporting to the registry or another error of data collection, we know from our daily interactions with patients and families that many people have concerns about vaccinating themselves or their children against COVID-19.

It is not a surprise that many people are hesitant about receiving the vaccine. It is new, though the technology for mRNA vaccines has been studied for decades. The rush to produce an effective vaccine quickly has made many people concerned that important safety steps have been sidestepped. However, because of the crippling effects of this global pandemic, the medical and scientific community responded in ways they usually don't:

(continued on page 2-3)

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
Ashley Mitchell-Ringuette, CCLS
Julie Sweet, BA, CF Research Coordinator
Schuyler Darcy, Medical Assistant
Kendra Arnold, Patient Service Specialist

ADULT PROGRAM
802-847-1158

Charlotte Teneback, MD, Adult CF Program Director
Zachary Weintraub, MD, Pulmonologist
Abe Sender, PA-C
Tara McCuin, PhD, Psychologist
Martine Antell, PharmD, Pharmacist
Christine Prior, LICSW, Program Coordinator
Kaitlin Steiert, RN, CF Nurse
Kitty Brady, RT
Maryann Ludlow, RD, CDE
Julie Sweet, BA, CF Research Coordinator
Katya Sajovec, Medical Assistant
Jenna Carroll, Patient Service Specialist
Susan Heney, Operations Support Specialist

COVID-19 Vaccine (continued)

- scientists switched from other projects to focus on vaccine development
- large amount of federal government money was funneled directly into vaccine development
- pharmaceutical companies started producing vaccines while trials were still ongoing so they could be ready soon after approval
- administrative barriers that could slow the process of approval down were removed.

The trials also did not take as much time to perform because given the uncontrolled spread of COVID in the community, the people in the trials were exposed to COVID-19 infection at higher rates than they would be if this was not a pandemic. All of these factors helped speed up the vaccine development and approval process without compromising safety.

The good news is that over 219 million Americans (including 14.8 million adolescents ages 12-17) have been vaccinated, providing even more evidence than the vaccine trials could that these vaccines are safe and effective. Even while case numbers have continued to go up, the rates of people who need to be hospitalized or who die from COVID-19 are lower in areas that have high vaccination rates.

Any vaccine can be expected to have some side effects that will show up immediately or up to a few weeks after vaccination. The side effects experienced have been rare and minor for most people who get the Pfizer BioNTech or Moderna vaccine and include pain at the injection site, fatigue, muscle aches, chills, and fever. These usually go away in a few days and are a sign that the immune system is working. However, vaccines have not been shown to have long-term effects that show up years later. The mRNA contents of the COVID-vaccine are broken down quickly by the body, so there is no biological explanation for how an mRNA-based vaccine would have long-term side effects. People are encouraged to report any symptom after vaccines to the Vaccine Adverse Event Reporting System (VAERS), which means that a lot of symptoms

(continued on page 3)

COVID-19 Vaccine (continued)

or events can be reported that had nothing to do with the vaccine. Vaccine safety experts review these reports to look for patterns or unusually high numbers of health problems so that possible vaccine-related side effects can be investigated. Sometimes this leads to actions like pausing the Janssen vaccine administration in April 2021 (which occurred after 6.8 million doses were administered and six people developed a rare type of blood clot). The VAERS reporting process can also unfortunately lead to claims that adverse events were vaccine-related when they were not, which is important to know whenever reading about possible vaccine side effects.

When we think about the possible risks of a vaccine, we often compare it to the risk of doing nothing and staying healthy, which makes it seem like choosing not to get vaccinated against COVID-19 could be a safer choice. But we can't lose sight of the fact that the risks of COVID-19 itself are more serious (and often more commonly reported) than the risks of the vaccine.

People with CF are considered to be at risk of developing serious illness if infected with COVID-19 because they almost always have some degree of underlying lung disease. The risk is higher for people with more advanced lung disease or those who have suppressed immune systems after having a transplant. The CF Foundation has been tracking closely the number of reported COVID cases in people enrolled in the CF registry since the start of the pandemic, and the reported rates for both children and adults are increasing 20-30% each week in recent weeks. Not all of these people will end up hospitalized, but hospitalizations are also on the rise - about 10% of patients with CF who get COVID have been hospitalized. For those who contract COVID but do not need to be hospitalized, the cost is measured in missed school, work, and activities, and placing others at risk. We also know when children contract COVID, it is often after an adult in their household contracts COVID. This makes it even more important for all household contacts of people with CF to be vaccinated.

At the UVM Cystic Fibrosis Center, 87% of eligible pediatric patients have been fully vaccinated against COVID-19, which is higher than the national average but still shows that some of our younger patients are not protected. 63% of eligible adult patients at our CF Center have been fully vaccinated, which is higher than national rates reported in the CF Registry but again, shows a large number of people vulnerable to complications from COVID infection. If you or your family members want to talk more about vaccination, please know that your CF care team is here to talk more with you about it.

Resources:

CFF Community Questions and answers:

https://www.cff.org/Life-With-CF/Daily-Life/Germs-and-Staying-Healthy/CF-and-Coronavirus/COVID-19-Community-Questions-and-Answers/

https://www.cdc.gov/coronavirus/2019-ncov/vaccines/facts.html

The Importance of Home Spirometer Use

By Kitty Brady, RRT

Before the pandemic, a Pulmonary Function Test (PFT) was standard with every appointment as it is a good way to monitor lung function and guide treatment. If your PFT numbers are dropping, maybe an increase in treatment is needed. Alternatively, PFT numbers can show if the treatment is working.

When the pandemic hit, people were told to stay indoors and to stay away from other people. This obviously made it very difficult to monitor your PFTs in the clinic. The Cystic Fibrosis Foundation (CFF) recognized this very early on and generously "bought" every person with cystic fibrosis (who wanted one) a home spirometer.

If you would like a home spirometer and have not yet received one, please let Mike Bissonette, RRT or Kitty Brady, RRT know and we will get one sent to you. Mike and I have taught everyone how to use the home spirometer properly. If we have missed someone or if you feel you need a refresher, please reach out to Mike or Kitty for training.

When you learned how to use the spirometer, we found your "baseline" on the home spirometer. In other words, we compared the Forced Exhalation Volume in 1 second (FEV1) from your Clinic FEV1 to your home spirometer FEV1. This way, if there were any differences between the two, we would keep this in mind when you sent in home spirometry results.

We encouraged you to practice using your home spirometer in order to watch your own FEV1. After training, we suggested you use it a few times a week for two weeks, then weekly for two weeks, then every two weeks thereafter (for pediatric patients) and monthly for adult patients.

Dr. Lahiri would like pediatric patients to send home spirometry results to the clinic every two weeks (either the 1st and the 15th or the 8th and the 28th), while the adult providers would like to see results monthly.

Both clinics would like to see results just before a clinic visit (remote or in person). We also ask that you use your home spirometer when you feel increasing symptoms. These include increased shortness of breath, increased cough, and increased fatigue. Call the clinic to report the new symptoms and your home spirometry results. Just like in clinic, when you come in sick and you have a PFT, the providers can use this information to make medical decisions.

Currently, we are asking everyone to please upload your results into your MyChart account and send them to one of the providers. If you are not sure how to do this, please reach out to either Mike or Kitty for help.

In the near future, you may not have to upload and send your results into clinic, as we are hoping to use the ZephyRx Dashboard. This will automatically upload your results into a provider dashboard where your providers can see them immediately. We will be training everyone on how to use the dashboard when we have worked out all the details. Stay tuned!

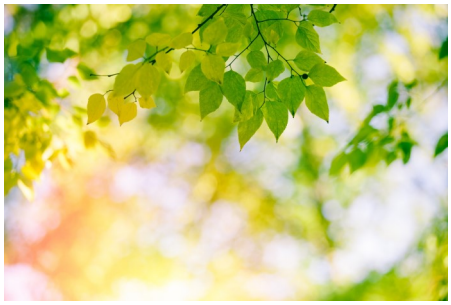
REDCAP PATIENT PRE-CLINIC SURVEY

By Emily Dischino, RN

The Pediatric CF Center will be instituting an electronic survey prior to your appointments. You will receive an email 1-2 weeks prior to clinic appointment. If your child is over the age of 15 years, we ask that both patient and parent each complete the survey. As for most families, we only have the guardian's email on file so if family is comfortable we would ask to also have the adolescent's email on file so they might also receive the survey link. We want to capture questions/concerns/topics that you wish to cover in your family member's upcoming visit. If you have any questions or concerns please don't hesitate to contact me at emily.dischino@uvmhealth.org or call our office 802-847-8600 to speak with a staff member.

Vitamin D Quality Initiative

By Maryanne Ludlow, RD, CDE



The UVMMC CF clinics have launched a new quality initiative (QI), jointly with the CF clinics at Maine Medical Center and Dartmouth Hitchcock Medical Center. This initiative aims to improve vitamin D levels in patients who have had low levels within the past six months on their current Vitamin D therapy. This project involves taking one or more doses of high dose (300,000 to 500,000 IU) vitamin D by mouth. This is given to patients at their clinic appointments. While this way of improving vitamin D levels is new for us, it's not a new therapy. It has been used at other CF centers, and it is safe to do.

If this therapy works to raise our patients' Vitamin D levels into the normal range, they will only have to take this extra Vitamin D does a few times a year in clinic, rather than have to remember to take Vitamin D daily or weekly.

We started this project in July. Along with the high dose of vitamin D, patients are given a survey asking them questions that focus on their thoughts about the Vitamin D that they currently take. Most of the patients who have returned the survey have indicated that a barrier to taking their vitamins is having to take a lot of pills, and also remembering to take them. Of the three fat soluble vitamin levels that we check, the Vitamin D level is the most likely to be low. So, we are hoping that addressing this with a high dose of Vitamin D a few times a year, given in clinic, will go a long way toward solving this chronic problem. It will also take the burden of remembering to take extra Vitamin D off of the patient.

Vitamin D has many roles in the body. While it is imperative to keeping bones strong, which aids in the prevention of CF related bone disease by maximizing absorption of calcium, that's not all it does. Adequate levels of Vitamin D in the body help control chronic inflammation. It also helps keep the immune system strong. Both of these roles are especially important in CF, which is a disease that involves chronic inflammation.

Likely because of these beneficial effects, people with adequate levels of Vitamin D have been shown to have lower incidences of certain cancers, such as colon cancer and breast cancer, and of heart disease. Recent studies have also shown a link between vitamin D deficiency and development of CF-related diabetes (CFRD).

Low levels of Vitamin D have also been associated with mood disorders such as depression. This is because of its role in regulating certain brain chemicals that influence mood.

So, you can see that the benefit of having Vitamin D in the normal range goes way beyond just keeping your bones strong!

If you are asked to participate in this initiative and you accept, you will be asked to bring a snack dose of enzymes, if you take them. We will supply a fat-containing snack in clinic. This is because all fat-soluble vitamins are best absorbed with a fat containing food and enzymes. The large vitamin D dose (called a "stoss" dose, as this is a German word for "push," as in a dose to push your level up) is given orally via six small gel caps for the 300,000 IU dose and 10 small gel caps for the 500,000 IU follow-up dose. Three months after your first dose we'll ask you to get a new Vitamin D level drawn, so we can see how well the dose worked. If your level remains low we will offer you a follow-up dose. This cycle will continue until a normal vitamin D level is obtained, or a year after your first dose. At the end of the year, we will provide you with another survey to find out about your experience with the dosing in clinic.

If this dosing has gotten your Vitamin D into the normal range, we'll continue to give you the high dose, once to several times a year, depending on how many doses it took to get you into the normal range in the first year.

We are hoping that this initiative will result in most if not all of our patients getting to and maintaining a normal Vitamin D level in the future!

Research Update

By Julie Sweet

Even during the ongoing COVID-19 pandemic, research aimed at advancing our understanding of cystic fibrosis (CF) and developing new treatments remains a top priority. There are many promising opportunities for people with CF to play an important role in helping to achieve these goals. For the first time, researchers are trying to understand whether people who are able to take highly effective therapies like Trikafta™ that treat the underlying cause of CF can safely stop taking some of their longstanding therapies. For people with CF who are unable to take these new therapies, we are deeply committed to research aimed at closing the gap and providing highly effective treatments targeting the root cause of CF to all. At the Vermont Lung Center, we are also currently involved in several research studies focusing on improving diagnosis and treatment of chronic lung infections affecting people with CF, such as those involving non-tuberculous mycobacteria (NTM) and *Pseudomonas aeruginosa*.

In addition to helping advance our understanding of CF, participation in a clinical trial can have benefits such as helping people with CF take an active role in managing their CF care and gain access to new treatments before they are more widely available. Although there are many benefits to participating in a clinical trial, there are also possible risks that can be serious. These risks include side effects of the treatments being studied, unwanted events during the trial that may or may not be related to the study treatment, and failure of a treatment to work. The decision to join a clinical trial is personal, and it is important to consider the benefits, risks, and time commitment required. We strongly encourage having conversations with trusted family, friends, and doctors and study coordinators to decide if a clinical trial is a good fit.

If you are interested in finding out more about current or future research studies, please contact the Vermont Lung Center for more information at (802) 847-2193. At this time, the studies below are open for enrollment:

SIMPLIFY

The SIMPLIFY study is being done to test whether or not it is safe to stop taking inhaled hypertonic saline or Pulmozyme® (dornase alfa) in people with CF that are also taking Trikafta™. As highly effective cystic fibrosis transmembrane conductance regulator (CFTR) modulator drug therapies like Trikafta become more commonly used, many in the CF Community (patients, families and caregivers) are asking if any of the other pre-existing therapies can be reduced or stopped. It is still largely unknown whether or not other chronic therapies can be safely stopped. The goal of the SIMPLIFY study is to get information about the safety of stopping either hypertonic saline or Pulmozyme by testing if there is a change in lung function in people with CF who are assigned to stop one of these medications as compared to those who are assigned to keep taking their chronic medication while continuing to take Trikafta. Participants who join the study will be randomly assigned to one of the following groups:

- Study A to keep taking hypertonic saline
- Study A to stop taking hypertonic saline
- Study B to keep taking Pulmozyme
- Study B to stop taking Pulmozyme

This study will involve 4 visits over about 8 weeks. Study visits may involve physical exams, measurement of lung function using spirometry, and paper and electronic questionnaires (done on your phone or tablet). Financial compensation provided up to $447.

(continued on page 7)

Research Update (continued)

This research study requires:

- Diagnosis of cystic fibrosis
- Ages 12 years and older
- Currently taking Trikafta™, as well as either hypertonic saline or Pulmozyme®, or both
- Willing to comply with the randomly assigned study treatment to either keep taking or stop taking hypertonic saline or Pulmozyme®, if applicable
- Willing to participate in 4 study visits over 8 weeks

PREDICT & PATIENCE

This is a research study to evaluate a standardized way of determining if nontuberculous mycobacteria (NTM) is responsible for worsening health (NTM disease) in people with CF who culture this type of bacteria in their lungs (Part A - PREDICT) and then evaluate a standardized approach to the treatment of NTM in people with CF with a diagnosis of NTM disease (Part B - PATIENCE). During the Part A - PREDICT portion of this study, participants will be seen for their regular CF clinic visits and no additional study visits are required. During the Part B - PATIENCE portion of the study, upon NTM disease diagnosis, participants will be asked to take antibiotics to treat NTM disease according to the study protocol and the Cystic Fibrosis Foundation Consensus Recommendations for Treatment of NTM Disease. Participants will continue to be seen for their regular CF clinic visits and no additional study visits are required for the study beyond those that are needed as part of the usual follow-up for this infection. Combined clinical care and research study visits for both PREDICT and PATIENCE parts may include collection of information about medical history and current health status, completion of questionnaires, and collection of blood, urine, and sputum (mucus) samples for the study. Financial compensation provided as follows: $60 enrollment visit, $15 per completed questionnaire, and $30 per research specimen collection.

This research study requires:

- Diagnosis of cystic fibrosis
- Ages 6 years and older
- History of at least one positive culture for non-tuberculous mycobacteria (NTM) in the lungs in the past 2 years

ABATE

The ABATE study is being done to test the safety and effectiveness of treating non-tuberculous mycobacteria (NTM) infections in people with cystic fibrosis (CF) with a drug called gallium nitrate. Previous laboratory tests suggest that this drug may be effective in combating NTM infections. Participants who meet all of the study requirements will receive two courses of 5-day intravenous (IV) infusions of gallium nitrate. The clinical trial will include a total of 8 study visits over about 5 months, with two of the visits lasting 7-10 hours each. Study visits will include questionnaires, diary completion, blood draws, urine and sputum (mucus) collection, and spirometry. Financial compensation provided up to $1620.

This research study requires:

- Diagnosis of cystic fibrosis
- Ages 18 years and older
- History of two consecutive positive cultures for non-tuberculous mycobacteria (NTM) in the lungs
- Willing and able to provide sputum (mucus) samples

(continued on page 8)

Research Update (continued)

Calithera Biosciences CX-280-202

This is a research study to learn more about the safety of a drug called CB-280 in people with CF. CB-280 is an investigational drug that is taken by mouth that blocks an enzyme called arginase, which is thought to play an important role in promoting lung infection in CF. This study is being done to identify which CB-280 doses are safe to take without causing too many side effects. Participants who meet all of the study requirements will receive either CB-280 or placebo to be taken by mouth twice daily for 14 days. The study will include a total of 6 visits over an 8-week period, with two of the visits lasting about 15 hours each. Study visits will include blood draws, physical exams, lung function testing such as spirometry, electrocardiograms, and sputum induction (collection of mucus/phlegm that is coughed up after breathing in saline). Financial compensation provided up to $1320.

This research study requires:

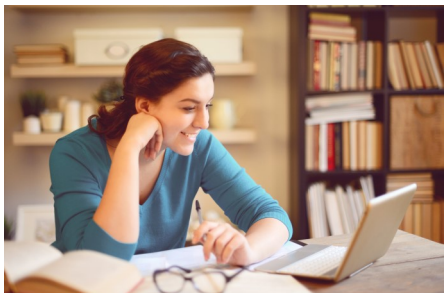
- Diagnosis of cystic fibrosis
- Ages 18 years and older
- Positive culture of Pseudomonas aeruginosa infection in the lungs
- FEV1 % predicted between 40 to 90%

LIMIT-NTM

This research study is being done to learn more about chronic lung infections involving non-tuberculous mycobacteria (NTM) and other lung microbes in people with CF by examining samples of sputum (mucus). Researchers are working to better understand what is happening in the lungs of people with CF who are chronically infected with NTM and how NTM infection interacts with other CF lung infection microbes. The information collected from this study may also help the researchers to develop better treatments for lung infections involving NTM. The study will include a single study visit during a CF clinical care visit at the University of Vermont Medical Center where participants will enroll in the study and provide a respiratory sample.

This research study requires:

- Diagnosis of cystic fibrosis
- Ages 18 years and older
- History of at least one positive culture for non-tuberculous mycobacteria (NTM) in the lungs
- Willing and able to provide a respiratory sample



CF Scholarships and Financial Aid

By Christine Prior, LICSW

Many scholarships and financial aid options are available for students with cystic fibrosis who want to pursue higher education. Below is a list of some of those. If you would like help finding additional 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.

CF Scholarship Programs AbbVie CF Scholarship Program
https://www.abbviecfscholarship.com/

Boomer Esiason Foundation (BEF) Scholarship Program
https://www.esiason.org/cf-living/scholarships

The Bonnell Foundation Marge Carmona Education Scholarship Program
http://thebonnellfoundation.org/scholarship/

Breathe for Bea Foundation Scholarship Program
https://www.breatheforbea.org/programs/scholarships/

Cystic Fibrosis Scholarship Foundation (CFSF)
http://cfscholarship.org/

Dana Walters Scholarship Foundation
http://www.dwscholarship.com/scholar.html

Elizabeth Nash Foundation Scholarship Program
http://www.elizabethnashfoundation.org/scholarships.html

The Living Breath Foundation Financial Aid/Academic Scholarship Program
http://thelivingbreathfoundation.com/aid.html

Susanna Delaurentis Charitable Foundation Scholarship Program
http://thesusannafoundation.org/scholarships/apply.php

United States Association of Cystic Fibrosis Adults (USACFA) Higher Education Scholarship (Lauren Melissa Kelley Award) and Scholarship for the Arts
https://www.cfroundtable.com/scholarships

THE VERMONT CYSTIC FIBROSIS SCHOLARSHIP

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 March 1, 2022. 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.

CF Foundation: Programs You May Not Have Heard Of

By Jim Gilbert, Development Director, CFF Northern New England Chapter

CF Peer Connect is a one-to-one peer mentoring program for adults with CF and their family members to connect about shared experiences. No matter what you're going through, there is someone who has been through a similar experience and can offer support. After you request a peer mentor, the CF Peer Connect team will match you with an adult with CF or a family member - parent, partner, or spouse - who has experience with the topics you want to learn more about.

LEARN MORE ABOUT CF PEER CONNECT

Community Voice is an empowering, virtual opportunity for people with cystic fibrosis and their family members to share their experiences, perspectives, priorities, and knowledge to impact CF research, care, and programs. We can't be successful without the input of the CF community, and Community Voice is the avenue for getting that input. Each person's experience is unique, so it's critical to hear from as many people as possible. Whether filling out a two-minute survey or participating in a focus group or committee, Community Voice members are helping to make a difference for every person affected by CF.

LEARN MORE ABOUT COMMUNITY VOICE

Effective September 1, 2021: We Have Moved.

Please Note Our New Address:

Cystic Fibrosis Foundation
Northern New England Chapter
20 Trafalgar Square, Suite 447
Nashua, NH 03063

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/

CDC:

https://www.cdc.gov/coronavirus/2019-nCoV/

VT Department of Health:

https://www.healthvermont.gov/response/infectious-disease/2019-novel-coronavirus