

THE
University of Vermont
 MEDICAL CENTER

THE
University of Vermont
 Children's Hospital

CF CONNECTION

Newsletter of the Vermont Cystic Fibrosis Center Advisory Board

EXERCISE IS MEDICINE



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Exercise Is Medicine

By Maggie Holt, Physical Therapist

To help CF patients get active and stay active, the CF Foundation is helping this center fund a physical therapist part time over the next three years. Maggie Holt will now have support for helping promote activity and exercise for our center's CF patients.

Our previous efforts to do annual physical therapy (PT) evaluations met with barriers around time, preparation and insurance coverage. With help from a CF Foundation grant, Maggie will have time to work on solutions to these barriers.

Why? Because, exercise is medicine. There is more than enough evidence to support that physical activity results in better lung function, fewer exacerbations and/or hospitalizations, better endurance and higher energy level for everyday life. Once you get into an exercise habit that's enjoyable, it doesn't feel like you're taking medicine - it feels like you're enjoying life.

There are countless options: train for a local road race, learn to rock climb, join a team, dance or do Zumba, go to the playground and play, ride a bicycle or a horse. There are thousands of ways to be active, and it's never too late or too early to start.

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VERMONT CYSTIC FIBROSIS CENTER

PEDIATRIC PROGRAM**802-847-8600**

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 Michael Bissonette, RT
 Carlie Geer, RD, CD
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 Julie Sweet, BA, CF Research Coordinator
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 Kitty Brady, RT
 Maryann Ludlow, RD, CDE
 Kristen Bolton, RD, CDE
 Christine Prior, LICSW
 Julie Sweet, BA, CF Research Coordinator
 Stacie Reed, Medical Assistant
 Lisa Benoure, Medical Assistant
 Jenna Carroll, Practice Support Specialist
 Susan Heney, Operations Support Specialist

New Faces in Clinic**Julie Sweet**

Clinical Research Coordinator

Julie is a native Vermonter and grew up spending her spring seasons working and smelling the sweet scents in the sugarhouse at her family's maple sugaring farm. She graduated from Middlebury College with a Bachelor's Degree in Molecular Biology and Biochemistry and earned high honors for her senior thesis project completed in the Spatafora Lab. After graduating, Julie worked for 10 years as the Senior Laboratory Technician in the Cipolla Lab at the UVM College of Medicine Department of Neurological Sciences. In her free time, Julie enjoys reading, playing with her 1-year-old daughter, Grace, and taking her yellow Labrador on long walks.

**Andrea Mucia**

Medical Assistant

Pediatric Program

**Kendra Therrien**

Practice Support Specialist

Pediatric Program



Dedication to the Memory of Donovan Couture

By the Vermont CF Center Advisory Board

It is with deep sadness that we dedicate this issue of *CF Connection* to the memory of Donovan Couture. Donovan passed away on December 31, 2016 at the age of 49. Donovan and his wife Jacie were founding members of the Vermont CF Center Advisory Board. Together, they were the seasoned veterans of our group, and we relied on their insight and experience to inform our work. We valued their perspective as patient and spouse, and their experience as parents navigating the challenge of explaining Donovan's hospitalizations, surgeries and treatments to their daughter Tiffany. For many of us in the CF community, Donovan was the first adult with CF that we had the opportunity to know. His resilience and positive attitude in the face of so many CF complications was inspiring. We will miss Donovan, and we offer our heartfelt condolences to Jacie and Tiffany.

Included below are the remarks Dr. Laurie Leclair made at Donovan's memorial service:

Fifteen years ago I arrived in Burlington to start a care program for adults with cystic fibrosis. I was taking over for a much-loved pediatrician and was often greeted with a healthy dose of skepticism as I met my new line up of patients. Who was I and what did I know about CF? I will never forget walking into Donovan's hospital room for the first time. He had had a lung transplant a few years earlier, and it didn't take long to appreciate that his history was complicated. I had no idea! As I met him and his beautiful wife Jacie for the first time, I was greeted with kindness, openness and respect. Clearly these two were a capable pair looking for a partner and advocate to ensure Donovan the best possible health care! What I didn't appreciate then, but know now, is that we were about to embark on one of the most important and fulfilling journeys of my medical career. Over the next 15 years, we navigated every health complication imaginable. And thanks to Donovan we did so with style and grace. There were surgeries, new medications, countless procedures and endless referrals. Not to mention trips to Boston and back. Through it all, he displayed nothing but a "get 'er done" attitude without a single complaint.

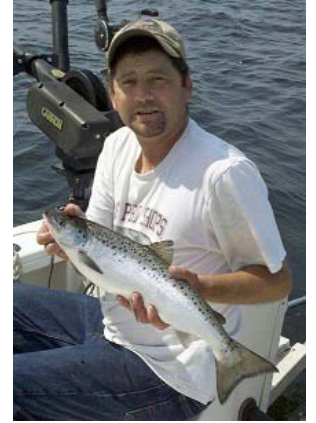
Donovan and Jacie were always frequent fliers in our CF

clinic but in recent years, they came nearly every week. We would try to take care of the medical piece quickly at these visits so we could spend time talking. I loved to hear about fishing or Tiffany's latest horse show or concert where she blew the crowd away. Donovan especially loved to tell those stories, and I especially loved to hear them as they represented a real "worth" to all the effort he put in to living.

Incredibly there has also been lots of laughter, even at times when things might not seem so funny. One such example I will never forget took place about 10 years ago, before his kidney transplant. Donovan had been on antibiotics for pneumonia at home and developed worsened kidney failure – uremia, as we call it. Jacie called and I told her to bring him in. When he arrived, Donovan could hardly wake up. I kept shaking him and asking "Donovan. Can you tell me my name?" After a couple minutes of this, Donovan picked his head up, looked at me and said "Dr. Whatever" and flopped back down. Jacie and I laughed until tears streamed down our faces, at a time when, frankly, all of us needed a good laugh. Dialysis and a kidney transplant later, Donovan was as good as new! And this story will always remain a favorite.

Over the years, Donovan and Jacie have done me lots of favors. They have been strong advocates for adults with CF amongst our patient and family group and were founding members. They have participated on regional panels to discuss the patient perspective of organ transplantation, and they have met with our medical students to share the family perspective of living with chronic illness. Through all of these efforts they have left a legacy, a real tangible story of what it means to live well with whatever hand you are dealt.

In many ways, I think Donovan gave me more than I provided to him in return. As we all try to fill the giant hole Donovan's loss has left in our hearts, I challenge you to live your life as he lived his. I challenge you to look adversity in the eye and keep going. And, most importantly, I challenge you to do so with a smile on your face and a kindness in your heart.





Exercise Is Medicine (continued from page 1)

What will Maggie's work with us look like for you? Over the next three years, you'll notice:

- Phone calls and emails between clinic visits
- Sometimes, activity monitoring (think Fitbits and pedometers)
- Sometimes, active challenges during clinic appointments (a chance to show off, or set a goal for yourself)
- Education about active airway-clearance techniques, chest expansion, core strengthening
- Activity ideas even for babies, preschoolers and their families
- A chance to sort out aches and pains in your muscles and joints if needed
- Referrals to PT close to home, if needed
- Ideas for other exercise opportunities in your community
- Another physical therapist to partner with Maggie so there's always someone available to help

The right amount of activity for kids is 60 minutes a day. Adults should get at least 150 minutes per week (300 preferred). This is meant to be "moderate-to-vigorous activity" (it should make you short of breath).

If something stands in the way of a healthy activity level for you, please talk to the team at your next clinic visit and we'll be in touch.

INPATIENT EXERCISE PROGRAM

By Abe Sender, PA

As many of you know, routine exercise is now being offered and encouraged as part of the inpatient treatment plan. Exercise is a very effective form of airway clearance, and sessions are taking the place of one treatment each day. Exercise sessions typically last about 30 minutes and are supervised and monitored by a respiratory therapist.

Patients have several options for type of exercise, including a treadmill, stationary bike, or exertional video games on the Xbox. Typically, exercise is performed in an exercise room, which is used for several CF patients per day. Strict contact precautions are maintained, similar to the process we use for exam rooms in CF clinic. Only one patient uses the room at a time and diligent cleaning occurs between patients. Some patients cannot use the room because of resistant bacteria. In these cases special exercise equipment is brought to the patient's room for regular exercise.

We have seen a significant interest in the exercise program and have had a lot of positive feedback.



Tara Morgan McCuin, PhD
Psychologist

Update on the CF Mental Health Grant

By Christine Prior, LICSW

In early 2016, the CF Foundation awarded our center a mental health grant. The grant allows both the pediatric and adult cystic fibrosis programs time and resources to better address mental health needs. The grant benefits our patients with CF and their caregivers. The CF Foundation's grant allowed UVM Medical Center to give me some protected time to create strategies that ensure improved access to quality mental health care for our patients. Here is some of what we have been working on so far:

- We created and distributed a survey to assess patient and caregiver's perceived and experienced barriers to accessing mental health care. If you have not yet completed this survey, please type the following address into your browser and share your experience:
<https://redcap.med.uvm.edu/surveys/?s=FKN9WMXLDN>
- We brought psychologist Tara McCuin on board as part of the CF team. Tara attends pediatric and adult inpatient rounds, meets with all newly diagnosed families and provides mental health consultation to the CF teams. She is an active member of the Vermont CF Center Advisory Group and sees a number of adult patients with CF for outpatient individual therapy.
- We continue with annual mental health screening in both pediatric and adult CF clinics.
- We are in the process of developing video clips to educate patients and caregivers on a variety of issues related to CF and mental health. The first of these has been filmed and is being edited so stay tuned!
- School outreach: Emily DiSchino, RN and I have presented to the special education directors for all the Chittenden County schools. We talked about the importance of offering mental health support to students in the school environment. I aim to offer this presentation to additional districts as well.
- We have begun outreach to county mental health agencies to provide education on cystic fibrosis and mental health. Also, we have begun to develop a network of providers who are informed on issues common for patients with CF. These providers are willing to collaborate with the CF team to provide comprehensive care. To date, I have been able to present to four county mental health agencies with two more scheduled in the next couple of months. These counties are in both Vermont and New York and are where roughly 100 of our patients live.

I am excited about this project and the work that has been done so far. I am hopeful that these efforts will result in easier access to quality mental health care for patient and families.

CF Foundation's Educational Webcasts

Paula Lomas, MAS, RN, CCRP, Director, Clinical Communications

Melissa Chin, Medical Programs Manager

In a continuous effort to improve your experience on [CFF.org](https://www.cff.org/) (<https://www.cff.org/>) we've made the CF Foundation's educational webcasts easier for you to find. You may now:

1. View the entire webcast collection under the [Playlists tab](https://www.youtube.com/user/CysticFibrosisUSA/playlists) (<https://www.youtube.com/user/CysticFibrosisUSA/playlists>) on the CFF YouTube channel.
2. View the webcasts as embedded videos within their respective content areas on [CFF.org](https://www.cff.org/). Example: "Partnering for Care" is embedded within "[Your CF Care Team](https://www.cff.org/Care/Your-CF-Care-Team/)" (<https://www.cff.org/Care/Your-CF-Care-Team/>)
3. View webcasts by topic using the following direct links:
 - [Respiratory](https://www.youtube.com/playlist?list=PLhoQ6vyZhqqo3qHxD2IsX6DCqTcinxghE) (<https://www.youtube.com/playlist?list=PLhoQ6vyZhqqo3qHxD2IsX6DCqTcinxghE>)
 - [Nutrition/GI](https://www.youtube.com/playlist?list=PLhoQ6vyZhqqo9F9-5ANhpr3H4MtSZudt) (<https://www.youtube.com/playlist?list=PLhoQ6vyZhqqo9F9-5ANhpr3H4MtSZudt>)
 - [Genetics](https://www.youtube.com/playlist?list=PLhoQ6vyZhqqosoC5cKpvAj0FOSzH2bsBy) (<https://www.youtube.com/playlist?list=PLhoQ6vyZhqqosoC5cKpvAj0FOSzH2bsBy>)
 - [Germs](https://www.youtube.com/playlist?list=PLhoQ6vyZhqqqs0rosMBCZFQBpY02yTAK4) (<https://www.youtube.com/playlist?list=PLhoQ6vyZhqqqs0rosMBCZFQBpY02yTAK4>)
 - [Transplant](https://www.youtube.com/playlist?list=PLhoQ6vyZhqqrVDmRx3R7JAYXS88PkHnVR) (<https://www.youtube.com/playlist?list=PLhoQ6vyZhqqrVDmRx3R7JAYXS88PkHnVR>)
 - [Age Specific](https://www.youtube.com/playlist?list=PLhoQ6vyZhqqrhV2gZRdo9y1N-1c8VC8q0) (<https://www.youtube.com/playlist?list=PLhoQ6vyZhqqrhV2gZRdo9y1N-1c8VC8q0>)
 - [Partnering With Your Care Team](https://www.youtube.com/playlist?list=PLhoQ6vyZhqqoepeiWPMNE2LqchpHK2N5t) (<https://www.youtube.com/playlist?list=PLhoQ6vyZhqqoepeiWPMNE2LqchpHK2N5t>)
 - [Research](https://www.youtube.com/playlist?list=PLhoQ6vyZhqqpE-0F1Qkiw_dt1gECFeBC) (https://www.youtube.com/playlist?list=PLhoQ6vyZhqqpE-0F1Qkiw_dt1gECFeBC)

These webcasts can also be viewed on tablets and smartphones. We hope you find this information helpful.

Reflection on "A Cure for All – Leaving No One Behind"

By Liz Hammel, Vermont CF Center Advisory Board

In October I went to the North American Cystic Fibrosis Conference (NACFC) in Orlando, Florida. It was the 30th time the Cystic Fibrosis Foundation has brought clinical staff from all disciplines (doctors, nurses, dietitians, respiratory therapists, and social workers) together with researchers, pharmacists, foundation executives and pharmaceutical reps from all over our continent and beyond. At the conference, those working the hardest on CF present their latest research, share information about quality improvement projects, announce initiatives and introduce new medications and therapies. CF clinical staff also meet by discipline to find out what some clinics are doing to get the best outcomes for their patients.

Overall, it is a scientific conference, so people like me, who are not doctors, don't understand a lot of the material presented in the breakout sessions. There are three times, though, when everyone at the conference gets together in what they call "plenary" sessions. This is when the Foundation celebrates how far CF care and research have come through the years. It also shares its vision of where it hopes we, as a CF community, will be in the near future.

This was my third trip to the NACFC and the first plenary session was the most hopeful one so far. It was called "A Cure for All – Leaving No One Behind." This session discussed that, while two medications are now available to correct CFTR (the basic defect that causes cystic fibrosis), those medications treat only approximately half of the people living with CF.

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Reflection on “A Cure for All – Leaving No One Behind” (continued from page 6)

Ivacaftor (Kalydeco) and the lumacaftor / ivacaftor combination (Orkambi) are not available for people who don't have certain mutations of CFTR. What the CFF means by “leaving no one behind” is that part of the new focus in CF research is something that will help all people with CF, no matter what mutation they have. This focus is gene repair.

It's difficult to describe something that I have only the most basic understanding of, but here's my best attempt. When the CFF talks of gene repair, they mean to not just focus on fixing the defective CFTR protein and making the chloride channel in the body's mucus-producing organs work correctly. Instead they are figuring out a way to make people with CF's bodies *make* normal CFTR in the first place. In other words, they are targeting the steps *before* the CFTR protein is made. This means fixing the DNA by breaking it at or near the defective gene. When the cell repairs the chromosome, they trick it into replacing the faulty sequence with the normal, or non-CF sequence. During the plenary session, the CFF had some very cool visuals that helped me picture how this would work at the cellular level, but no description of mine will compare.

The take away is that, because this happens *before* the CFTR protein is made, this gene therapy will be available to all with CF, regardless of their mutation. Why? Because it won't matter in which way their CFTR malfunctions (or if any is made to begin with, or if enough is made, or if it might be trapped in the cell), because the CFTR will be *made* correctly. There has been success in the laboratory in this type of gene editing. Apparently, this breaking of DNA and subsequent gene repair has been done in the lab.

The next challenge is taking what is done in the lab and getting it to the body. How do we get the repaired DNA into the body and into the nucleus of the correct cells? I have no idea. What I realized at the conference, though, and what gave me hope, is that there are some brilliant people dedicating their life's work to figuring it out. And what they are doing is working on a cure for *all* people living with CF.

Lung Transplantation

By Charlotte Teneback, MD

Lung transplantation is an emotional topic in many ways. It means recognizing the presence of end-stage lung disease and the risk of death. Also, it means recognizing that transplanted lungs are a gift from someone who has died. At the same time it is also somewhat of a medical miracle. For the last 25 years, lung transplantation has been a source of hope and added years for thousands of individuals.

The timing of when to begin the discussion about lung transplantation is complicated. Because the average survival after transplantation is relatively short (6.5 years in people with CF, though we have patients living 10-15 years and more after transplant), it is important that patients not receive a transplant before they really need it. On the other hand, patients need to be strong enough to handle the transplantation process. This can be difficult if done when their disease is very advanced. We typically bring up the topic of lung transplant with patients who have had a recent rapid decline (such as a big increase in how often they are ill or a rapid drop in lung function), have very poor lung function (FEV1 < 30%), or who have started to need oxygen or other breathing support.

Because there is no lung transplant center in Vermont, the first formal step of the process is referral to an out-of-state center, usually in Boston. Boston Children's, Brigham and Women's, and Massachusetts General hospitals all perform lung transplants. Prior to the first visit, a provider from the UVM Medical Center CF team will send a summary letter along with clinical information to the transplant center. This includes results of CT scans, lung function, bacterial cultures, and other information. The initial visit can then be scheduled, usually within one to two months.

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Lung Transplantation (continued from page 7)

At the first visit to the transplant center, the patient will meet with a physician (usually a pulmonologist, but sometimes a surgeon) as well as a coordinator. A significant amount of education is provided at this visit, so it is important to bring family and other key supporters along. The visit can take a full day, and families sometimes decide to spend the night in Boston on the day before or after the visit.

Based on the first visit, the transplant team will make an assessment about the appropriateness of transplantation. There are several different possible outcomes. They may feel someone is appropriate, but too healthy, so they will not do any testing—but they will ask to see the patient back in three-to-six months. They may not offer transplantation because they feel that there are too many barriers that cannot be overcome; these could be medical and/or social barriers.

There are many different reasons why someone may not be appropriate. These include obesity, failure of other organs such as kidneys, certain infections such as HIV or hepatitis, lack of social support, and addiction. Of note, no tobacco exposure at all is acceptable, including second-hand smoke exposure and the use of nicotine gum. These factors are considered barriers primarily because people who were transplanted under similar circumstances in the past did very poorly.

If a candidate appears appropriate and there are no significant barriers, the transplant team offers further testing. This is an essential part of the pre-transplant work-up. If the patient and family, and the transplant center, are in agreement to proceed, an inpatient stay for testing is scheduled. This inpatient stay takes place at the transplant center and may include tests like a cardiac catheterization, endoscopy (a video exam) of the stomach, ultrasound of the heart, and CT scan of the lungs.

There are also additional consultations with a psychiatrist, social worker, infectious disease physician, dietician, physical therapist and other specialists. There may also be additional testing or procedures that need to take place closer to home. These may include dental work, vaccinations, and physical therapy or pulmonary rehabilitation.

Of course, there are lots of insurance details that need to be worked out as well. While medical insurance covers transplantation, many families also fundraise to cover out-of-pocket expenses, such as staying in a local hotel and time off of work. Social workers at the transplant center will help guide this process. Overall, the testing process can stretch over three-to-six months.

Once all the testing is reviewed, the transplant team will make a final decision about listing. Being listed means that someone is on the transplant registry and is available as a match for donated organs. Once someone is listed, they can be called at any time and must be prepared to rapidly travel to the transplant center with short notice. There are resources available to help with this, including options for medical air travel if needed.

The wait time for transplant is variable between centers and can range from days to many months. Wait time depends not only on how ill someone is, but also on their body size, blood type, and availability of organs. Some very exciting work has been done in the past several years to increase the number of lungs available for transplant. Lungs are often damaged by the process that injured the donor, but new methods are being developed that are able to give lungs time to heal so that they can ultimately be used instead of discarded.

What can you do while you wait for all this to happen? There are some really important things that improve the chance of being listed and of successful transplantation. Maintaining a good nutritional status and staying active are incredibly important. Most transplant centers require participation in pulmonary rehabilitation during the transplant process. And of course, keeping in close touch with your local CF team to maintain the best health possible is critical.

Cystic Fibrosis Screening Guidelines

The following list contains screening guidelines for individuals with cystic fibrosis. These are recommendations from the Cystic Fibrosis Foundation to monitor and maintain health for people with CF.

Pediatric Screening

By Tom Lahiri, MD

Laboratory/Blood work

- Annual labs to look at electrolytes, liver function, kidney function, blood counts and vitamin levels
- Low vitamin levels may need a dose change and a recheck in 2-3 months
- Infants usually have their first set of labs within 1-2 months of starting CF vitamins
- Patients on Orkambi (lumacaftor/ivacaftor) need liver tests checked every three months for the first year
- For children who are taking high dose ibuprofen (or Motrin), levels need to be checked every two years or when there has been a 25 percent weight increase. These level checks take three hours

Radiology

- Annual chest x-ray for most children
- Chest CT scans can be done in place of x-rays and be performed at 1-3 year intervals, depending on the findings and the child's lung health
- Bone density studies are not routinely indicated for pediatric patients
- Sinus CT scans are sometimes done in patients with chronic sinus disease, particularly if surgery is being considered

Respiratory cultures

- Throat or sputum cultures are done at each visit if one or months apart
- Sputum for atypical Mycobacteria (AFB) should be done 1-2 times per year in people who take chronic azithromycin

Visits

- Infants are seen monthly for the first year
- For children over 1 year of age, quarterly visits are recommended with sooner follow up as needed for either nutritional or pulmonary issues
- All children should have at least annual visits by a nurse, respiratory therapist, dietician and social worker

Diabetes

- All children with CF should be screened for CF-related diabetes annually, beginning at age 10

Immunizations

- Annual influenza vaccine is indicated for all infants and children 6 months and over
- Routine pediatric immunizations (e.g. DTaP, Hib, HepB, PCV, MMR) should be administered according to recommended schedule at the pediatrician's office
- PCV-13 booster should be given to children who have not received this
- PPSV-23 should be given to all children with CF over 2 years of age with a booster every 5 years

Eye exams

- Annual eye exams are recommended for children who are taking Orkambi
- Eye exams are indicated for children with insulin-dependent CF related diabetes after 5 years of treatment

Depression/Anxiety screening

- Annual depression and anxiety screening for patients 12 years of age and older
- Annual depression and anxiety screening of parents and patient caregivers

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Adult Screening (continued from page 9)

By Laurie Leclair, MD

Laboratory/Blood work

- Annual labs to look at electrolytes, liver function, kidney function, blood counts and vitamin levels
- Low vitamin levels may need a dose change and a recheck in 2-3 months
- Patients on Orkambi (lumicaftor/ivacaftor) need liver tests checked every three months for the first year
- For adults who are taking high dose ibuprofen (or Motrin), levels may need to be checked every two years or when there has been a 25 percent weight change. These level checks take three hours.
- Urinalysis for microalbuminuria in all patients with CF-related diabetes

Radiology

- Annual chest x-ray
- Chest CT scans may also be performed depending on how patients is doing
- Bone density study every five years after age 18, more frequently (1-2 years) if on chronic steroids or have abnormal baseline
- Sinus CT scans are sometimes done in patients with chronic sinus disease, particularly if surgery is being considered

Respiratory cultures

- Throat or sputum cultures are done at each visit
- Sputum for atypical Mycobacteria (AFB) should be done 1-2 times per year in people who take chronic azithromycin

Visits

- Quarterly visits are recommended with sooner follow up as needed based on how the patient is doing overall
- All patients should have at least annual visits by a nurse, respiratory therapist, dietician and social worker

Diabetes

- All patients with CF should be screened for CF-related diabetes annually with an oral glucose tolerance test
- For patients with CF-related diabetes, a hemoglobin A1C should be performed quarterly
- A minimum of quarterly endocrinologist follow-up is also recommended

Immunizations

- Annual influenza vaccine is indicated for all patients
- PCV-13 booster should be given to all patients who have not received this
- PPSV-23 should be given to all patients (6-12 months after the PCV13), with a booster every five years until 65 years of age

Eye exams

- Eye exams are indicated for patients with insulin dependent CF-related diabetes

Depression/Anxiety screening

- Annual depression and anxiety screening for patients and caregivers

Patient Perspective: Lung Transplantation

By Kate Heffernan

I knew that lung transplantation had always been an “end of the road”-type treatment for cystic fibrosis patients, but I didn’t want to believe I was nearing the end of my road. I remember the day I was sitting in my hospital bed waiting for my doctor to come talk to me about my recent health decline. I couldn’t tell you the month - maybe it was in February. I couldn’t tell you the day, or even the time, but I could tell you every feeling and thought that was going through my mind before she came in and told me what I had been fearing for the entire hospital stay. My pulmonologist, who I had been seeing since I turned 18, walked into my room. After a short discussion about recent changes in my health, she told me that it was time to consider looking into lung transplantation.

She left my room that day and I remember feeling like I had failed my body and everybody that loved me. I tried so hard to stay healthy so that I could pursue my dreams. Prior to this hospital admission, I had been studying veterinary medicine at the Royal Veterinary College in London, England. During the end of my second year, I fell ill and flew home for medical treatment. After the four-week hospital stay in February, admissions became more frequent, and I began using oxygen. At first it was for exertion only. The next hospital stay was a short two months after my discharge in March, and this stay wasn’t very long. After this, there was more oxygen and pulmonary rehab, but the situation still did not seem so dire. I continued to work part time and be active. It would be the following admission that changed the game – and for real this time.

That September, about six months after my last hospital stay, I was admitted into the Intensive Care Unit through the Emergency Department on a Saturday morning. There, in the ICU at The University of Vermont Medical Center is where I sat for almost six weeks, where I struggled nearly every day, and where I would begin testing to be considered for the transplant list.

Between these two stays, we made a trip down to Boston to meet with the Lung Transplant team at Massachusetts General Hospital. In July of 2014, seven of my family members and I drove down to stay the night before beginning an early morning at Mass General. The day began at 8 am, and I had an appointment to meet with everyone on the team in the clinic that day. I met the nurses, the doctors, the support staff, the nutritionist, the social worker, the psychologist and the financial advisor. All of it was a little overwhelming, but we got through it and we left the hospital with a lot more information about the whole process than I can even remember to this day.

The stay in the ICU in September would be the beginning of all my testing to be considered for the transplant list. Normally they like to do all the testing down in Boston, but given my current situation - being very ill and on a very high amount of oxygen at rest - Boston gave the okay for me to start my testing at UVM Medical Center. For two-or-three days straight, I felt a little bit like a lab rat, with all the poking and prodding. I can’t remember what test they decided to start with but there were many - a right-heart cardiac catheter, a gastric emptying study, a swallow study, a ventilation/perfusion study, a six-minute walk test, pulmonary function tests, echocardiogram, and I’m sure there were more. If there weren’t more, it sure felt like there were, because this was the sickest I had been up to this day. I required a lot of treatments and testing during this hospital stay that I had not yet experienced. Then, after all my information was sent to Boston, all we could do was sit and wait for any further instruction.

In December I got a call from Mass General telling me that I would be put on the active lung transplant list, if I would like to be. Being on the list and then getting the transplant is all up to the patient. I was told I could stop the process at any time, except when I am under anesthesia receiving lungs. I agreed, and the waiting process began. I knew I would have to have my phone on me at all times, in case *the* call came through saying there was a match. I also would make a trip down to Boston every three months so that they could monitor changes in my health, in between my visits at UVM Medical Center.



Kate kayaking on her birthday, post-transplant

Patient Perspective: Lung Transplantation (continued from page 11)

Once I was told I was on the list my family and friends jumped on fundraising as much as they could to help with any costs associated with my transplant journey. They held a dance and did many bake sales and product fundraisers. They even made sweatshirts in support for my journey and had a variety show with a silent auction – all to help me. My community came together, and I cannot even believe how much support I received through my whole journey – from people who may not have even known my family or me.

For nearly a year, I went down to Boston every three months or so when I wasn't in the hospital. Through this time, I needed oxygen for everything. I was mostly home bound, and if I wasn't, I had to pack up oxygen tanks to bring with me for every visit, short or long.

Then, almost a year after being listed on the active transplant list, I got THE call. I had just gotten out of the hospital, on a Friday after a three- or four-week stay. It was a Tuesday and I was sitting at home eating lunch and watching TV when my cell phone rang. It was Mass General. Not thinking anything of it, I answered the phone and began chatting with the transplant nurse, like usual. Then she began asking me if I had eaten anything today. That is when I knew something was up. She told me they were looking into some things, not to eat anything more, and to wait for her to call me back in a half hour or so. The next half hour passed so slowly. I went up to my room and packed a few things and tried to remain calm. I put on my vest and did a treatment. The transplant nurse then called back and told me to get down to Boston as soon as I could. She said not to rush because they were still waiting on a couple things, but get to Boston as quickly and safely as possible. I had so many calls to make, but the most important was to find someone who was around to drive me there. I tried my mother, who was at work and would take about forty-five minutes to get home. I tried my sister; same boat. Then I called my father. He was right in town and was able to get his things together quickly to come pick me up and head down.

Once we got down there at round seven or eight in the evening, I was admitted to the hospital. They hooked me up to an IV, drew a bunch of blood, and told me to sit tight for any information about the lungs being a match. My hospital room was full. Several family members drove down in anticipation for the upcoming news and hopefully life-saving surgery. Most of them stayed the night in my room and the others searched for a hotel room to stay in after getting updates. Around midnight, we got the news that if it was “a go” I would go into surgery around four or five in the morning. Occasionally, transplant recipients will get THE call and get to their transplant hospital, only to find out the lungs aren't a perfect match. This is what they call a “dry run.” There was part of me that was hoping this might be the case because I felt like I wasn't ready for my life to change so drastically. But I knew I had the support and that I could do it. I had no other choice.

It was a fairly restless night, and when the team was headed back to the hospital after the organ procurement, I was told the lungs looked good and that I would begin preparing for surgery around five in the morning. I was terrified, but I was ready. That morning I was wheeled down for surgical prep, said goodbye to my family, and wouldn't awake to see them again for another 24 hours.

After surgery, I was intubated and had all kinds of lines for all different things coming out of me. There were four chest tubes, a couple IVs, an arterial line, a jugular catheter, a urinary catheter, an epidural, oxygen on for recovery, EKG lines and an oximeter. Once I was extubated, I woke up in a glass box (really a room in the surgical ICU) with people around me in isolation gowns, masks and gloves - all to protect my newly immune-compromised body. Shortly after waking, I was encouraged to get up and go for a walk. I thought they were crazy, but I did it. I knew I had to. It was time to start my recovery.

Patient Perspective: Lung Transplantation (continued from page 12)

As the days went by, seemingly very slowly, things started to improve. Lines and tubes began being pulled, and for the first time in years, I was free of my oxygen. The usual stay in the surgical ICU (SICU) is three-to-five days depending on progress. Due to some other factors my stay in the SICU was about a week long. I was making progress, but one of the bugs that came from my donor prolonged my stay. I was walking daily and began to work with physical therapy at least once daily while in the SICU. When it was time to make the move to the step down unit the only things I still had attached to me were a single chest tube and my PICC line. I stayed in the step down unit for about five days, where it was a little less like living in a glass box. It was more of a normal hospital room. Visitors still had to wear a gown and gloves, but it was much more normal than the previous few days.

I was discharged from the hospital on a Friday. I was happy but also very nervous. I had my mother to stay with me and help me with whatever I needed, but this was her first experience with transplant too, so neither of us knew what to expect. Not having medical professionals just outside my room to ease my mind about what is normal and what is not made me very uneasy - but we did it and we never looked back.

We rented an apartment about a five-minute walk from the hospital as we would need to make frequent trips back for the first four-to-six weeks. I settled into my room at the apartment and took a moment to sit and take everything in, not knowing what to think of the immediate future. For the next few weeks, I had two or three appointments every week. After that, I had one or two appointments a week. I had blood work done, a chest x-ray taken and then met with the transplant team at least once a week. Blood work was done until the medication reached the desired level in my system. Until then there were a lot of medication adjustments. Eventually, I started doing pulmonary function tests again, which I was very nervous to do with my new lungs, as well as one-on-one physical therapy two-to-three times a week. While I was there, I also had appointments to meet with many different teams and get additional post-transplant testing done. These procedures/tests vary from patient to patient depending on their individual experiences.

My first trip home was in December and after that I got to come home for a week for the holidays. Then I spent one last week down in Boston after the New Year to finish up appointments, and I headed home for good after that. For the following year, I would make monthly trips down to Boston - sometimes day trips, sometimes overnights if I had to have a procedure done the day after my transplant clinic. Back home in Vermont, I began one-on-one physical therapy as there was no pulmonary rehab program available at the time. Over six weeks, I became so much stronger and learned a lot about using my new lungs and body. During my physical therapy and shortly after, I became an avid hiker and started back to work almost full time.

At my year anniversary for transplant, I went to Boston for a couple nights because there are yearly tests that need to be done. They are basically the ones that I had pre-transplant and then shortly post-transplant to compare to my baseline and monitor for any changes. These tests include bronchoscopies to check for any rejection and any organisms that may be growing down in the new lungs. After my year checkup, I was told that my appointments would now be every three months! My bronchoscopy looked great, my pulmonary function was at its highest since I can remember, and I was doing very well.

I still have to be careful about certain things, as you would expect because I am immune-compromised more than the normal CF'er, but I have never been able to breathe like this. I have never laughed so hard without having a huge coughing fit after. I have never been so grateful for every single person in my life. Donate Life encourages people to write to their donor family. I haven't quite figured out the words to say to express my and my family's gratitude for their loved one's amazing gift that he gave to me. I am back to work and am able to go out without having to plan each move beforehand. I am living the most normal and amazing life. I was always skeptical and unsure if I got to the point of needing transplant if I would go through with it. Now I know it was the right choice, and I would do it over again because I was not ready to end what had just barely started.

CF Resource Corner

By Jennifer Eddy, CCLS

Welcome back to the CF Resource Corner! We have been working hard to ensure that patients and families are up-to-date on current age-appropriate educational materials and helpful resources. Watch as the list grows over time. The newest resources are marked with an asterisk. How many of the resources below do you have? And how many have you browsed or used recently? Please feel free to reach out if you are missing one or more. They are meant to be tools for caregivers, siblings, teachers, nurses and others. We can provide extra copies if needed.



Welcome Bag

- Patient and Family Checklist
- *Beginning CF Care Handbook*
- *Who I Am* book
- *Milestones in CF Care: Newly Diagnosed/Early Childhood*, book 1 of 3
- Salt Administration Handout
- Child Life Specialist Flyer

***CFLifeLessons.com Video Clip** - A five-minute motivational video about a young adult with CF reflecting upon her desire to have done a better job with treatments and medications. Provided to ages 12 and above.

***Medication Bag** - A neoprene bag to hold medications (minus those that need refrigeration) that will be brought back and forth to each clinic visit for medication reconciliation. Provided to every family with a child newborn and older. Feel free to decorate. See Child Life for Sharpies.

***Weekly Pill Organizer** - A box with 28 individual compartments to fill with medications. Provided to every family with a child newborn and older. Made possible by UVM Children's Hospital and Children's Miracle Network Hospitals.

***MyHealth Online** - A secure patient portal offering you a quick and easy way to manage your health. You can view lab results, growth charts and immunizations, request prescription medication renewals, message your doctor's office, pay medical bills and more. Child Life can help you get set up right in clinic!

NEW PHARMACY SUPPORT COMING!

By Charlotte Teneback, MD

The UVM Medical Center's CF Center was chosen for a grant from the CF Foundation to support part-time pharmacy support for both the pediatric and adult clinics. In the coming months, a pharmacist will join the rest of the team for clinic visits. Also, pharmacy technicians will assist with prior authorizations, refills and similar issues. The pharmacists will meet with patients and their families to review medications, interactions and side effects. They will also help us ensure appropriate monitoring where indicated. We are hopeful that this will help us improve care in many ways, including reducing wait times for medications and ensuring appropriate use of all medications.

Current Clinical Research at the Vermont Lung Center

By Julie Sweet, Clinical Research Coordinator at the Vermont Lung Center

This is a very exciting time in cystic fibrosis clinical research! There are many promising opportunities to help in the development of new treatments. These range from therapies to improve quality of life by treating the symptoms of CF, to treatments that are even targeting the cause of CF. At UVM Medical Center, one study is of antibiotics to treat lung infections and CF exacerbations. Another is a study to improve nutrition and growth in children with CF. Besides helping advance our understanding of CF, participation in a clinical trial can have direct benefits: one benefit is helping people with CF take an active role in managing their CF care; another is that participants gain access to new treatments before they are more widely available. Although there are many potential 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. It is important to consider the benefits, risks and time commitment required. We strongly encourage having conversations with trusted family, friends and doctors to decide if a trial is a good fit.

Novartis CTBM00002407

This is a five-year, observational study in CF patients with the chronic respiratory infection *Pseudomonas aeruginosa* (*P. aeruginosa*) treated with TOBI Podhaler or another FDA-approved inhaled antipseudomonal antibiotic drug. The study sponsor, Novartis, is conducting this study to look at how well TOBI Podhaler or other FDA-approved inhaled antibiotic medications work on treating the bacteria in the lungs of CF patients. It is also looking at their long-term safety. Researchers will test the effectiveness of these drugs by tracking lung function and the amount of *P. aeruginosa* in sputum collected during regularly scheduled clinic visits. They will also track pulmonary exacerbations and other measures.

Inclusion Criteria:

- Diagnosis of cystic fibrosis
- Ages 6 years or older
- Established diagnosis of *P. aeruginosa* lung infection, having at least two documented positive *P. aeruginosa* cultures in the last year (may include first visit)
- Prescribed and initiated chronic treatment with FDA-approved inhaled antipseudomonal antibiotic medication for treating *P. aeruginosa*
- Actively enrolled or willing to enroll in the Cystic Fibrosis Foundation registry
- Willing to attend at least 2-3 regularly scheduled, routine clinic visits

Exclusion Criteria:

- Documented FEV1 less than 25% in the previous year
- Current participation in an interventional clinical study with an inhaled antibiotic treatment

STOP2

Pulmonary exacerbations are treated with varying antibiotics for varying time periods based on individual needs determined by patients, their families and care providers. Both doctors and CF patients have questions about the best way to treat pulmonary exacerbations. Doctors and CF patients want to make sure the patient gets antibiotics long enough to get better but not so long as to expose the patient to unnecessary risks. This study is for people with CF who experience a pulmonary exacerbation and are planning to receive IV antibiotic treatment. This study will look at the safety and effectiveness of three different lengths of IV antibiotic treatment for pulmonary exacerbations. Participants who respond early to IV treatment will receive antibiotics for 10 or 14 days. Those who do not respond early will receive antibiotics for 14 or 21 days. Researchers will study the effectiveness of the different IV treatment lengths by measuring changes in lung function. This study requires lung function tests and/or other measurements. Financial compensation provided up to \$210.

Current Clinical Research at the Vermont Lung Center (continued from page 15)

Inclusion Criteria:

- Diagnosis of cystic fibrosis
- Ages 18 years or older
- Actively enrolled in the Cystic Fibrosis Foundation registry
- Doctor's confirmed plan to initiate IV antibiotics for a pulmonary exacerbation
- Willing to perform spirometry and answer short questionnaires during all three fixed visits at the beginning, middle and end of the study
- Willing to adhere to a specific treatment duration determined by initial response to antibiotic treatment and then randomization
- Willing to return for follow up visit after treatment has completed

Exclusion Criteria:

- Treatment with IV antibiotics in the six weeks prior to enrollment
- Primary diagnosis for this hospitalization is unrelated to worsening lower respiratory symptoms (for example, sinusitis)
- History of solid organ transplant

GROW (Enrolling in 2017)

Nutritional study for pancreatic-insufficient children with CF with the goal of improving weight gain. Enrollment is planned to begin in 2017.

March on the Hill: It's Not Political, It's Personal

By Laurie Eddy

Every year the CF Foundation recruits advocacy volunteers from each state to come to Washington, DC for the Annual Cystic Fibrosis March on the Hill. It is a chance for CF advocates to share their personal stories with their Congressional representatives while advocating for the CFF's policy agenda. This will be the 11th year for the March on the Hill and I will represent Vermont for the first time.



One of the most important aspects of the meetings with our members of Congress and their staff is telling our personal story. I will tell stories, share photographs and give our members of Congress a clear picture of the struggle our family faces in the fight against CF. Having a loved one with CF is a unique experience. Sharing our personal story enables us to act not only as an advocate but also as an ambassador for those with the disease. With all that's changing in health care at this moment, it's critical for our members of Congress to know the reality of living with and paying for CF care.

Significant steps have been taken toward repealing the Affordable Care Act. As critical health care reform decisions are made in Washington, it is essential that members of Congress hear from people in the CF community about the health care coverage needs of people with cystic fibrosis. I will ask our members of Congress to keep our CF community's needs in mind while they work to build a health care system that will benefit all Americans:

1. We need assurance that any health care reform will support a system that provides adequate, affordable and available health insurance for people with CF.
2. We need robust, reliable funding for biomedical research at the NIH to support a strong pipeline of CF therapies and a well-resourced FDA to ensure new treatments are safe and effective.

You can follow the March on the Hill on social media. Follow #CFAdvocacy on Instagram and Twitter to see what volunteers across the country are doing to advocate on behalf of those with CF.

CF Scholarships and Financial Aid

The Vermont Cystic Fibrosis Scholarship

A Vermont family donates this \$500 scholarship.

Applications are due by April 15, 2017. To apply, email Christine Prior, LICSW a letter stating your interest to Christine.Prior@UVMHealth.org. Recipient's name will be drawn at random from qualifying candidates. Eligible patients are those students who have not received this scholarship previously and are enrolled in a college program for the fall.

The Cystic Fibrosis Scholarship Foundation

The Cystic Fibrosis Scholarship Foundation (CFSF) is a scholarship program for students with CF enrolling in an undergraduate college program or a vocational school in the fall. CFSF was founded by a parent of a young adult with CF and is neither part of nor funded by the Cystic Fibrosis Foundation.

Scholarships of \$1,000 each are awarded and may be used for tuition, books, and room and board, for up to one year of study. Students may reapply the following year for an additional award, but are not guaranteed future awards. Awards are sent directly to the institution that the student is attending. Scholarships are awarded based on a combination of financial need, academic achievement and leadership. For application forms, deadlines and other information, visit www.cfscholarship.org or call 847-328-0127.

AbbVie CF Scholarship

Applicants include US citizens with CF who are enrolled in, or awaiting acceptance from, an accredited institution in the fall. Awards are based on applicants' creativity, academic excellence, community involvement and ability to serve as a positive role model for the CF community.

Forty scholars will be selected based on their academic record and extracurricular activities, essay and creative presentation. Each selected student will receive a \$2,500 scholarship. Of these forty recipients, one undergraduate and one graduate student will receive a Thriving Student Scholarship of more than \$20,000. The application and additional information about the scholarship can be found at www.AbbVieCFScholarship.com/apply.

The Elizabeth Nash Foundation Scholarship Program

The Elizabeth Nash Foundation Scholarship Program awards scholarships to assist persons with CF to pursue undergraduate and graduate degrees. Scholarships are awarded on the basis of demonstrated need and accomplishment, both academic and other. Grants are made each year to people with CF who exhibit clear academic goals and a commitment to participate in activities outside the classroom. Grants are made directly to the academic institution to assist in covering the cost of tuition and fees. For application deadlines and more information, visit www.elizabethnashfoundation.org.

Other Resources

For other options, contact the George Washington University HEATH Resource Center for a publication entitled, "Creating Options: Financial Aid for Students with Disabilities," which provides contact information and background about financial assistance opportunities. To obtain a copy, please contact:

The George Washington University HEATH Resource Center

2121 K Street, NW, Suite 220

Washington, D.C. 20037

Phone: 202-973-0904

Fax: 202-994-3365

E-mail: askheath@gwu.edu

www.heath.gwu.edu

Federal Student Aid Information Center at the U.S. Department of Education provides a publication entitled "The Student Guide" which lists many financial assistance programs:

Federal Student Aid Information Center

P.O. Box 84

Washington, DC 20044-0084

Phone: 800-4-FED-AID (800-433-3243)

www.studentaid.ed.gov

POSTAGE
HERE

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.

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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

- **Link to Dr. Beall's Presentation**

Dr. Beall, former President and CEO of the Cystic Fibrosis Foundation, spoke at the VT CF Center Advisory Board's last education event in the fall. We are pleased that we can share Dr. Beall's presentation with patients and with those caregivers unable to attend. Here is the link to the CF Advisory YouTube channel video of Dr. Beall's talk entitled "Can we do better?"

[Dr. Beall's Presentation \(https://www.youtube.com/watch?v=jaxSOH5wZeQ&feature=youtu.be\)](https://www.youtube.com/watch?v=jaxSOH5wZeQ&feature=youtu.be) Video by Myles Jewell

- **CF Caregiver Support Groups**

Cystic Fibrosis Research, Inc. in California offers caregiver support groups for parents of children with CF and parents, spouses or partners of adults with CF. Participation is open nationwide by phone. For more information, please call (855) 237-4669 or go to: <http://www.cfri.org/supportgroup.shtml>