



THE  
**University of Vermont**  
 MEDICAL CENTER

THE  
**University of Vermont**  
 Children's Hospital

## CF CONNECTION

Newsletter of the Vermont Cystic Fibrosis Center Advisory Board

### A SENSE OF EMPOWERMENT AND HOPE



*Kitty Brady RT, Derek Hammel, Liz Hammel, Carol Baker, Christine Prior LICSW, and Martine Antell PharmD attend a plenary session at the NACFC*

## North American CF Conference

By Carol Baker

My husband and I were given the honor of representing the Vermont CF Center Advisory Board at the North American CF Conference in the fall. We are the parents of Emma, a 16-year-old living with CF. Although the conference was overwhelming at times, it was an empowering experience that left us with a feeling of hope. Everyone associated with CF has felt times of depression, helplessness and fear. After attending the conference, those feelings have become a little easier to handle.

The moment we walked into the conference center was amazing. To know that every person in attendance is working for the same cause was very emotional for me. And then to learn that the cause is not just for better treatments and care, but a cure for ALL those suffering with CF, filled us with a huge sense of hope.

One of the sessions I attended was titled "CF Interventions Advancing through the Clinical Testing Phase." Although I did not understand much of the medical lingo, the purpose of the session was to review some of the latest trials of CF therapies including both CFTR modulators and anti-inflammatory

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## VERMONT CYSTIC FIBROSIS CENTER

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 Jillian Sullivan, MD, Associate CF Program Director  
 Keith Robinson, MD, Pediatric Pulmonologist  
 Tara McCuin, PhD, Psychologist  
 Martine Antell, PharmD, Pharmacist  
 Julie Houchell, FNP  
 Emily DiSchino, RN, CF Nurse Coordinator  
 Melissa Barron, RN, Pulmonary Nurse Coordinator  
 Kelly Little, RN, Pulmonary Nurse  
 Heather Coons, RN, Pulmonary Nurse  
 Michael Bissonette, RT  
 Carlie Geer, RD, CD  
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**ADULT PROGRAM****802-847-1158**

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 Tara McCuin, PhD, Psychologist  
 Martine Antell, PharmD, Pharmacist  
 Abe Sender, PA-C  
 Erick Duprey, RN  
 Kitty Brady, RT  
 Maryann Ludlow, RD, CDE  
 Sasha Morey, PT  
 Christine Prior, LICSW  
 Julie Sweet, BA, CF Research Coordinator  
 Allyson Airolidi, Medical Assistant  
 Lisa Benoure, Medical Assistant  
 Jenna Carroll, Practice Support Specialist  
 Susan Heney, Operations Support Specialist

**Food Insecurity Screening****By Keith Robinson, MD**

Being healthy requires a network of support in our schools, workplaces and community. Regular access to food is an essential part of becoming healthy and maintaining it. It may surprise you that roughly 13% of households in the United States experience food insecurity. This means that over one in eight families have trouble affording food during the year.

Access to healthy food is critical for patients and families with cystic fibrosis because adequate nutrition is essential to promote growth, lung health and fight infections.

The UVM Children's Hospital Cystic Fibrosis Center will begin confidentially screening families for food insecurity so we can increase access to food resources. The screening will be done when children are admitted to the hospital and at the quarterly clinic visits. Parents will be asked two questions on a paper screening tool. Our team will work with families in need to find local resources to increase access to food.

Please feel free to contact us with questions. Our goal is to promote the health of our community with cystic fibrosis.

## North American CF Conference (continued from page 1)

therapy. The use of ivacaftor (Kalydeco) and the combination of ivacaftor with lumacaftor (Orkambi) has been very effective for some patients. This session addressed the more difficult-to-treat mutations. Researchers are combining different combinations of ivacaftor with other medications, including tezacaftor. Overall it has been well tolerated and the results are promising. They discussed the trial of tezacaftor/ivacaftor on patients with one copy of deltaF508 and one nonsense mutation. I was extremely excited when one of the slides showed Emma's exact mutation. Unfortunately, it did not show how effective the new medication was in addressing her mutation because it was mixed in with a bunch of others. But, just to know that her specific mutation is being studied gave me a great sense of optimism. There were relatively low adverse effects and no increase in respiratory events for patients in the study. There is a CF patient out there with Emma's same mutations who is taking a medication that could potentially change her life! These results were very exciting and it still gives me goosebumps when I think about it.

The second half of this session was dedicated to anti-inflammatory medications. This interested me in particular because Emma has been taking ibuprofen for several years. I believed she was about to reach the age where she would be taken off of it. High doses of ibuprofen are given to patients to keep inflammation in the lungs to a minimum. When inflammation is minimal there is less chance of infection settling in the lungs. Inflammation begins early in life and, when it is excessive, it accounts for irreversible lung damage. However, less than 5% of eligible patients are prescribed ibuprofen. I was pleased to hear that it is not always necessary to end this treatment around age 18. I believe it has been a major reason why Emma's lungs have had such little decline for the past several years.

We learned a great deal at this conference and were very pleased to be able to attend and represent Vermont. There were many highlights. We met some wonderful people including parents, clinicians, and Nick Talbot (a CF patient). We met Nick at a CF Connect dinner for families. He raises money for CF research through climbing mountains. He is currently climbing Aconcagua, the highest peak in South America. His goal is to climb the highest peak on each continent. He has reached four of the seven already, including Mt. Everest. Meeting Nick and the others can help make an otherwise lonely disease become a little less lonely. At the NACFC, seeing the amount of optimism, the research on so many different aspects of the disease, the number of people who share the same goal, and the end game of a cure for all who suffer from CF made a huge impact on my life.

### PEDIATRIC CLINIC TO USE TEXT MESSAGING



The Pediatric CF Clinic plans to attempt to use alternate forms of communication to remind patients and families of upcoming appointments. We have found that it is often difficult to reach most patients by phone and usually have to leave a message. We rarely receive return calls. Similarly, automated messages sent to home phone numbers are rarely answered. Many physician and dentist offices have transitioned to the use of text messaging as their primary means of communication with patients. In the coming weeks or months, the clinic will try to send text message reminders of upcoming appointments in addition to the usual reminder call. The patient or parent will be able to respond once to the text. Of course, the ability to proceed with this project will depend on the accuracy of mobile/cell numbers that we have on file. Please bear with us as we try to find a more effective way of communicating with you!

## CF in College

by Will Corcoran



*Will at the Botanical Gardens in Washington, DC*

My name is Will Corcoran and I am a 20-year-old CF patient attending the University of Vermont. Transitioning to college wasn't easy. In my third year, I am still figuring out how I can best manage my treatments, schoolwork, exercise and social life. Thankfully, UVM has worked out to be a great fit for me. Some challenges I have faced as a CF patient in college are: fitting in all of my treatments, taking responsibility to order medications, communicating with professors about my illness, and keeping my dorm clean.

When looking at colleges, my parents and I decided that whatever school I chose needed to be close enough to a CF center that I can get there easily in emergencies or when I'm not well. While living on campus the CF center was a ten-minute walk away. The convenience

made it easy for me to quickly go in to get seen by a doctor or pick up medications. Fitting in all of my treatments every day isn't easy; I am often going back and forth from campus to home. It is important when creating your schedule to think about when you will be able to do medications during the day and when your clinic's hours are so that you have that time free.

Eating in college is not easy and is something I am still figuring out. I find that having snacks at home and on me at all times is helpful, and exploring all dining options on campus is a good way to find multiple ways to get the food you need. Dining hall food is repetitive so switching it up so that you're always eating enough is a good plan.

When I left for college, I also left my CF clinic and support system behind. It's important to find a provider within your new CF clinic who works well with you so you stay well. When you move off to college, keep your support system with you. I am always keeping my parents in the loop about my health so that we can make decisions together. When I first got to school, I was hesitant to share my CF with new friends, but as I opened up to them I realized how accepting everyone is of my illness. Don't be afraid to be open with your new peers as they will soon become part of your support system as well.

Through speaking with other CF patients, I found that exercise seems to be one of the hardest things to manage at school. It took me until my junior year to finally get a good routine down. Looking back, I wish I had focused more on making time for exercise. I often block off times, as you do for class or treatments that are dedicated to working out in any way. Sometimes I will go to the gym (be sure to wipe down equipment); other times I will ski or go for a run. Nowadays, I have a personal trainer, which is very helpful. Most colleges have student personal trainers who are either very cheap or free, as they are learning too. This may be a great option for you.

School and my CF care often collide, but I've found that being up front with my professors and communicating often is the best way to go about it. Professors appreciate it when students are up front and honest. Also, speaking with them one-on-one early in the semester makes them remember you all year. Be sure to work with your Students with Disabilities office often so that you can get proper accommodations. There should be no reason you are ever penalized for late work or absences because you are sick.

In my third year at school, I am still learning everyday how to best manage my CF. In the end, if you make your health your top priority and surround yourself with family and friends that help you to stay well, you can do great at school. I love speaking with other CF patients and families, so please feel free to reach out to me at [willcorcoran20@gmail.com](mailto:willcorcoran20@gmail.com). Wishing you all a happy and healthy 2018!

*Vermont Cystic Fibrosis Center Advisory Board invites you to*

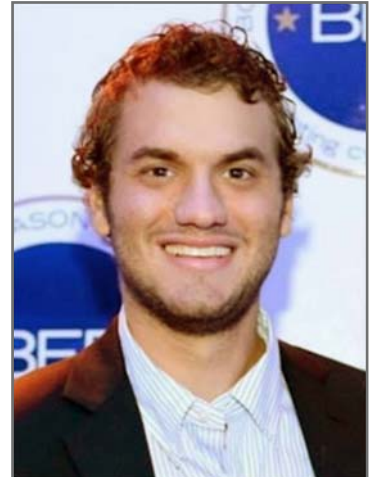
***The 10th Annual***  
**Patient & Family Education Night**

**Friday, May 4, 2018**

**5:30 – 9 pm**

**Champlain Room, Champlain College  
 Burlington, VT**

Gunnar Esiason is 26 years old and has cystic fibrosis. After being diagnosed at 2 years old, his parents founded the Boomer Esiason Foundation. Since then Gunnar has grown to live his life despite CF. He played three varsity sports in high school, graduated from Boston College in four years and now works as a cystic fibrosis patient advocate with the very foundation his parents started. He is also the head coach of his high school alma mater's varsity ice hockey team, making him the youngest head coach on Long Island. Gunnar has learned that success with cystic fibrosis is possible, and he hopes to share that with his audience tonight!



**Agenda**

5:30 - 6:00 pm	Registration, mingle with other families, and visit vendor tables
6:00 - 7:00 pm	Dinner and presentation of CF Center data by Tom Lahiri, MD and Laurie Leclair, MD
7:00 - 7:30 pm	Mingle with other families and visit vendor tables
7:30 - 9:00 pm	Keynote

**Registration is required. Go to <https://survey.uvm.edu/index.php/473626>**

**Deadline: Friday, April 27, 2018**

**This free event includes dinner and is open to caregivers, professionals, friends and relatives of those affected by CF.**

In keeping with the CFF Infection Prevention and Control Policy, only one person with cystic fibrosis may attend an indoor event sponsored by a CF Center. That person will be our guest speaker, Gunnar Esiason. We invite people with CF to view the recording of this event on our YouTube channel (video will be posted after the event).

## New Faces in CF Clinic



### **Martine Antell, Pharmacist for Pediatric and Adult Clinics**

Last summer, Martine began working with the CF Clinics. She had recently returned home to Vermont after some time away. She lived in Minneapolis, MN for five years and worked at the University of Minnesota Masonic Children's Hospital as an inpatient pharmacist. Most recently, she spent a year in San Francisco where her husband was completing a fellowship. She took this opportunity to stay home with their two young children. While there she continued to work with her colleagues at the University of Minnesota researching pediatric medication indications. Since Martine filled the CF clinical pharmacy position at UVM Medical Center, she has loved providing this medication-focused service. Martine is excited to be a reachable resource for families who need assistance or have questions about their medications.



### **Sasha Morey, Physical Therapist for Adult Clinic**

With a grant from the CF Foundation, Sasha Morey has joined the adult CF team on both the inpatient and outpatient services. Sasha is originally from Cincinnati, Ohio. She moved to Vermont for college, ultimately earning her Doctorate of Physical Therapy from UVM. After graduate school, she worked in a private practice outpatient clinic. Her emphasis was on return to sport and orthopedic and chronic injuries. Now she works full time at UVM Medical Center in Acute Care with an expanding focus on pulmonary rehab. Sasha hopes to bring a broad range of skills to the adult CF programs. She will perform yearly physical evaluations of patients within the clinic; she is also a resource to help with orthopedic/chronic injuries, exercise prescription, and improving exercise tolerance. Sasha looks forward to being a resource for any of your physical therapy needs.

## Tezacaftor/Ivacaftor Combination Drug Approved

The Food and Drug Administration has approved the tezacaftor/ivacaftor combination drug Symdeko for people with cystic fibrosis ages 12 and older who have two copies of the F508del mutation OR one of 26 specified mutations, even if they do not have a copy of the F508del mutation. In the near future we will reach out directly to those of you who are eligible for this therapy to discuss this exciting opportunity. We believe this medication will be available to patients in the near future but have not yet been notified of a specific timeline.

## 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.

There are many types of scholarships available based on different criteria. For example, scholarships might be based on involvement 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.](#) They maintain 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 the state and national scholarships available, UVM Medical Center offers **The Vermont Cystic Fibrosis Scholarship**. This \$500 scholarship is donated by a Vermont family and patients with CF are invited to apply. Applications are due by **April 1, 2018**.

To apply, email Christine Prior, LICSW a letter stating your interest at [Christine.prior@uvmhealth.org](mailto:Christine.prior@uvmhealth.org). The recipient's name will be drawn 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 Resource Corner

By Jennifer Eddy, CCLS



### RESOURCES INCLUDE:

- Welcome Bag
- Medication Bag
- Weekly Pill Organizer
- Patient and Family Checklist
- “Beginning CF Care” Handbook
- “Who I Am” Book
- “Milestones in CF Care: Newly Diagnosed/Early Childhood” book 1 of 3
- Child Life Specialist Flyer
- My Health Online Brochure
- CFLifeLessons.com Video Clip
- Living with CF Series
- Huxi: A Coloring Adventure
- An Adult Coloring Activity

Welcome back to the CF Resource Corner! We have been working hard to ensure that patients and families are up-to-date on age-appropriate educational materials and helpful resources. Watch as the list grows over time. The newest resources are **bolded**. 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, etc. We can provide extra copies if needed.

**Parenting with Love & Logic Handout** - The Love and Logic Institute has created a two-page summary of the popular *Parenting with Love and Logic* book. It offers tips about parenting styles, responsibility and guiding children to solve their own problems. Also, covered are consequences vs. punishment, encouraging realistic choices, helpful one-liners and miscellaneous tips. This resource is given to families at age two.

**9 Tips for Teaching Kids Responsibility** - Parents of young children (2 years old) are provided with this resource created by Care.com. It offers useful tips to help parents encourage responsibility early on in life. Reviews modeling, praising, managing expectations, avoiding rewards, providing structure and routine, and teaching consequences. “Responsibility is an attitude, the idea of taking action and being proud of doing it, not just always having your caregiver do it for you.”

**CF Voice.com** - An online community where people of all ages can benefit from motivation, inspiration and connection. Educational content includes games, art, cartoons, an interactive laboratory, videos, podcasts and articles. The resource is given to families at age 6.

**Team Impact** - Nationwide nonprofit organization that matches up families and local sports teams in order to help children and teens gain strength, camaraderie and support while teaching student athletes about courage, resiliency and life perspective. Children 5-15 are eligible. You may self-refer or a CF team member can help you with the process.





From left, Liz and Derek Hammel and Carol and Will Baker at a CF Connect Dinner at the NACFC

## NACFC: a Parent's Perspective

By Derek Hammel

In November, I attended the North American Cystic Fibrosis Conference (NACFC). As the parent of a child with CF, I was inspired by all the great information shared and by the cutting-edge research on display. The NACFC highlighted all the hard work so many people are doing to enhance the lives of all those with CF and their work towards a cure. Over three days there were many sessions to choose from, spanning a wide range of topics. It was a bit overwhelming at times! The best news I heard was that the predicted median life expectancy for someone with CF (per the CFF Patient Registry) increased to 47.7 years in 2016, compared to 33.9 years in 2001. That is amazing progress.

One session that I wanted to highlight was the Friday plenary session Lung Transplantation: Challenges & Opportunities for Advanced CF Lung Disease:

[https://www.nacfconference.org/Content/Archives/2017/Plenary\\_Session\\_II\\_Friday,\\_November\\_3,\\_2017/](https://www.nacfconference.org/Content/Archives/2017/Plenary_Session_II_Friday,_November_3,_2017/)

This session was the first time a plenary was devoted to lung transplantation. Although this is a difficult topic, lung transplantation is a reality for many people with CF. Because CF lung disease is progressive, transplant is sometimes considered as a treatment for advanced lung disease. The goal is to prolong and improve quality of life. This session reviewed the progression of lung disease and the option of lung transplant to treat advanced disease.

The first CF lung transplant was performed in Toronto in 1984. Since the late 1980s, over 4,000 annual CF lung transplants have been performed. More than half were done in the United States. In 2016, 265 individuals with CF in the U.S. received lung transplants. Post-transplant survival rates are unpredictable (ranging from days to decades), but have improved greatly since the 1990s. The CFF has recently developed a Lung Transplant Initiative. It is a multi-pronged approach emphasizing coordination between the CF Center team and the CF transplant team to develop best practices for lung transplant referrals.

This plenary session included several powerful short videos about people with CF (or not) who have successfully undergone lung transplantation. The process they went through was referred to as a "journey." The journey includes the stages of referral, evaluation, listing, and transplant. Everyone's experience is unique, and it was interesting to hear stories from Mindy, Charity and others.

Mindy was diagnosed with CF at birth and lives in North Carolina with her husband and daughter. Soon after moving to Raleigh, she got extremely sick and had an FEV1 of only 37%. After several hospitalizations and many consultations, her new CF care team brought up the topic of transplantation. Mindy described the discussions as "scary." However, she was deeply humbled and impressed by the community of caregivers who walked down this road with her. They looked beyond just her medical charts and factored in the strong support network she

*Continued on page 10*

## NACFC: a Parent's Perspective (continued from page 9)

had, as well as her own personal resilience. They treated her case as the unique situation it was and communicated clearly and thoughtfully with Mindy and her family. She was added to the transplant list in November 2009 and received a double lung transplant in January 2010. Happily, she is once again living a healthy, full life.

Charity Tillemann-Dick is an American-born soprano and top-selling classical recording artist. She is also a recipient of two double lung transplants. Charity was diagnosed with Idiopathic Pulmonary Arterial Hypertension in 2004. In September 2009, she received a double lung transplant at the Cleveland Clinic in Ohio. After complications from rejection, she had a second double lung transplant in January 2012. The main take away from Charity's story was how important it is to have the difficult but critically important conversations about advanced lung disease, transplantation and even death. Here is what she said:

"One of the responsibilities of doctors is to help you understand what the different paths forward look like. And to do that with delicacy and with kindness, with concern and with thoughtfulness. And it seems like a particularly difficult job to ask of people who've trained to keep their fellow earthly sojourners alive. Ask, 'What do we want this to look like?'... It's a really important conversation that we each need to have with each other, and with our doctors, because death happens. And delaying those conversations doesn't help anybody."

Since receiving her second transplant, Charity has continued her singing career. She also shares her story and vocal talents at numerous conferences and events around the country.

The journey of lung transplantation is a roller-coaster ride with its ups, downs, twists, and turns. By starting the tough conversations early and having a strong support group to help individuals go through the emotions mentioned (denial, fear, shock, anxiety, hope, acceptance, and relief) it seems that lung transplantation can be a transformational, life-enhancing procedure.

SAVE  
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DATE

# 2018

## Virtual Events for the CF Community

### CF MiniCon: Self-Care and Relationships

FEBRUARY 27 *Registration opens January 31*

*For adults with CF and their families*

### CF MiniCon: Sexual and Reproductive Health

APRIL 8 *Registration opens March 12*

*For adults with CF*

### CF FamilyCon

JUNE 3 *Registration opens May 1*

*For adults with CF and their families*

### CF MiniCon: Transplant

AUGUST 5 *Registration opens July 2*

*For adults with CF and their families*

### BreatheCon

SEPTEMBER 28-29 *Registration opens August 24*

*For adults with CF*

### CF MiniCon: CF Adults Connect

NOVEMBER 7 *Registration opens October 10*

*For adults with CF*

*\*All events are for people 18 and older*

POSTAGE  
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## 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

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## Online Resources

- The Vermont CF Center Advisory is now on Twitter. Follow [@VTCFAdvisory](#) for the latest Tweets about what's going on in the CF community. Keep up to date on local, regional and national CF news and announcements.
- [Cystic Fibrosis News Today](#): 27 Hacks That Can Improve Life with Cystic Fibrosis
- Share your unique insights and perspectives with the CF Foundation. Join Community Voice and help drive improvements that will benefit everyone affected by CF. Email [communityvoice@cff.org](mailto:communityvoice@cff.org) for more information. Follow Community Voice on Twitter [#CFvoice](#)
- Cystic-fibrosis-related diabetes is common for people with CF. Learn more about how it is treated. <https://www.cff.org/Life-With-CF/Daily-Life/Cystic-Fibrosis-Related-Diabetes/?linkId=42174608>
- Want to help plan cystic fibrosis fundraisers in your community? Contact your local chapter to get involved. [no-new-eng@cff.org](mailto:no-new-eng@cff.org)