

THE  
**University of Vermont**  
 MEDICAL CENTER

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 Children's Hospital

# CF CONNECTION

Newsletter of the Vermont Cystic Fibrosis Center Advisory Board



ROOFTOP GARDENING

## Gardening Opportunity for Adult Patients with CF During Their Hospital Admissions

By Lisa Hoare

As a Vermont Certified Horticulturist working at the UVM Medical Center, I occasionally have the unique opportunity to share with patients my love and appreciation for working with plants. Most often, this takes place in the Rooftop Garden at the Main Campus. Sometimes it's a patient experiencing the joyful feelings of wind and sunshine for the first time in a few weeks. Other times it's any of our smaller visitors who insist on visiting the garden as a special treat to make those frequent but necessary trips to the hospital more fun!

Most recently, I've enjoyed working with a few adult patients living with cystic fibrosis who have come to the Rooftop Garden to share in fun tasks such as harvesting chamomile flowers for drying or picking flowers for the tables in the new Garden Atrium. Time in the garden can help provide patients a moment or two to step outside of themselves for a while. They experience the calm that comes from being surrounded by the numerous smells, colors and sounds always present in the garden. Glimpsing a goldfinch snacking on sunflower seeds or the many busy bees helping pollinate zucchini plants offers a simple happiness. Watching nature provide for itself nurtures us. It provides a comforting reminder that we are all a part of the same community.

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**Gardening Opportunity (continued)**

Garden Time is available on Thursdays from 12-2 pm. If you are an adult patient with CF and would like to take advantage of this opportunity during your admission, please let Christine Prior know and she will schedule a time with me. We will be gardening until the end of the September and then resume again in the spring.

**CF Resource Corner****By Jennifer Eddy, CCLS**

Welcome to the CF Resource Corner. This is a place for sharing summaries about the resources provided to you by your CF Team. Each quarter, certain resources will be highlighted as a way to encourage you to read or re-read valuable information in your spare time. As children grow and develop, new ideas and strategies to maintain health and happiness are often helpful. This is because needs, priorities and behaviors change with time. Information can be found in a number of ways these days - verbally, through handouts, books, websites, online forums, newsletters and education nights. This column will focus on a wide range of tools.

The first highlighted resource is the “welcome bag.” Currently at diagnosis, new families joining the CF clinic receive a welcome bag. It includes a variety of resources that reinforce information shared verbally. New resources will be handed out over time as we receive them. Please feel free to reach out if you are missing one or more of the resources below. They are meant to be tools for caregivers, siblings, teachers, nurses, etc. We can provide extra copies if needed.

Welcome bag includes:

- Patient and Family Checklist - Visual schedule for Wednesday clinics with questions about physical and emotional health and a fun activity on the back.
- *Beginning CF Care* Handbook - Booklet about CF with glossary of terms, helpful worksheets and action plans.
- *Who I Am* Book - Early school age level book with illustrations that promote the importance of routines with treatments, enzymes, meals/snacks, exercise, school and play. *(continued on page 8)*

## QUIT SMOKING LINKS



For help with quitting smoking,  
please go to:

- Vermont: [802quits.org](http://802quits.org)
- New York: [NYSmokefree.com](http://NYSmokefree.com)

## Smoking and Cystic Fibrosis

By Tom Lahiri, MD

People with cystic fibrosis (CF) usually develop lung disease related to abnormal, thicker mucus in their airways. This leads to inflammation and infection in the airways. In people without CF, mucus is carried up and out of the airways by small hair cells, called cilia, which move in unison. In CF, however, the thick mucus lacks water, so the cilia become flattened and don't work correctly. This makes it easier for mucus to stay in the bronchial tubes and can lead to more irritation and infection.

Cigarette smoke is unhealthy for the CF lung. In a large study called the EPIC trial, it was shown that kids with CF who grew up with smoking parents were more likely to have lower lung function by the time they were 6 years old, compared to kids whose parents did not smoke. This negative effect was even seen in infants with CF who had increased evidence of lung inflammation and earlier lung disease.

Exposure to tobacco smoke in the home environment also increases the need for asthma medication in children, including those with CF. There are adolescents and adults with CF who smoke. This can lead to even more significant complications, including pulmonary exacerbations (or flare-ups that may require hospitalization), lower lung function and permanent damage to their lungs. Tobacco smoke can directly injure the cilia and lining cells of the airway.

We have definitely heard from some patients with CF that they smoke marijuana and that they are not worried because it is "safer." Smoking marijuana is not safe for people with CF and leads to increased cough, sputum production and wheezing. While lung function does not drop as much compared to tobacco smokers in the short term, there is an increased chance of developing a pneumothorax, or popped lung, which can be life threatening.

In our own CF center, smoking is still a big problem. In 2015, 41 percent of our pediatric patients were exposed to cigarette smoking in the home environment, either by a parent or another caregiver. This is MUCH higher than the national average of 17 percent. While smoking outside of the house is better than smoking inside, parents can still bring cigarette smoke into the home on hair, clothing, etc. In kids whose parents smoke "outside," we can still measure levels of nicotine by-products in their urine.

Our center will continue to ask patients and families about smoking. We understand that nicotine addiction is a very difficult problem to overcome. There are state programs in both Vermont and New York (Quit Lines) that can assist patients and families with tools for smoking cessation. CF Center staff can also help, so please let us try!

Vermont Cystic Fibrosis Center Advisory Board invites you to

## Save the Date: Educational Event for the CF Community

**Saturday, October 15, 2016 9:00 – 11:30 a.m.**

UVM Medical Center, Burlington, VT



### Keynote Speaker:

**Robert J. Beall, PhD**

Former President & CEO of the  
Cystic Fibrosis Foundation

Under Dr. Beall's leadership, the CF Foundation achieved unprecedented gains in research, treatment and care, and as a result, people with CF are living longer, healthier lives than ever before.

## AGENDA

9:00 – 10:00	Keynote address by Robert J. Beall, Ph.D.
10:00 – 10:15	Break with refreshments provided
10:15 – 11:30	Roundtable discussions on topics of interest to the CF community

Registration is required. To register, go to [https://  
www.surveymonkey.com/r/CFEventOct15](https://www.surveymonkey.com/r/CFEventOct15)

**Deadline: October 7th**

To help reduce the risk of cross-infection, we will be following the CF Foundation Attendance Policy for this event: <https://www.cff.org/About-Us/About-the-Cystic-Fibrosis-Foundation/Attendance-Policy-for-CF-Foundation-Events/>

This **free** event is open to caregivers, professionals, friends and relatives of those affected by cystic fibrosis.

## Keeping Your Nebulizer Up to Date and Clean

A clean, updated and properly working nebulizer system at home is essential to effective home therapy. Every three years it is possible to get a new nebulizer machine; please let our office know if you have an old one and would like us to reorder a new machine for you. Old machines can take longer to get treatments done and replacing them could help in cutting down your treatment time.

Keeping your nebulizer cups cleaned daily will ensure you are getting the full benefit of your treatment as well as cutting down on bacterial growth on your equipment. Please see below on acceptable ways to clean your nebulizer cups. Your local equipment vendor can make monthly deliveries of neb cups, if not every other month. If you haven't been receiving regular deliveries of these nebulizer cups, please contact Emily Dischino, nurse coordinator, and she will happily get you set up with these deliveries.



*Respironic Sidestream nebulizer*



*Pari LC Sprint nebulizer*

### HOW SHOULD I CLEAN MY NEBULIZER?

1. Wash your hands.
2. Use a paper towel to wash the inside and outside of the nebulizer parts with liquid soap and hot water. Throw out the paper towel.
3. Rinse the nebulizer parts with water.
4. Air-dry nebulizer parts on a clean paper towel or clean dish towel.

Once the nebulizer is cleaned, it can be disinfected. Remember to clean the nebulizer right after you use it, otherwise the medication can dry and harden making it difficult to clean.

### HOW SHOULD I DISINFECT MY NEBULIZER?

Although vinegar was recommended in the past, vinegar should not be used to clean/disinfect the nebulizer because it is not strong enough to kill the germs a person with CF has. Be sure to read the package insert for cleaning instructions, and if allowed, use one of the disinfecting options below:

#### **Boil for five minutes.**

For safety reasons, it is recommended that you not leave the nebulizer parts boiling unattended. Use an audible timer to remind yourself.

Air-dry the nebulizer parts by placing them on a clean paper towel and covering them with another clean paper towel. You should replace the paper towels with dry ones when they become wet until the parts are fully dry. This will help to stop the spread and growth of germs. Once dry, the parts should be stored in a dry, clean bag.

#### **Use the dishwasher, if the water is 158 degrees F or more for 30 minutes.**

If necessary, air-dry nebulizer parts by placing them on a clean paper towel and covering them with another clean paper towel. You should replace the paper towels with dry ones when they become wet until the parts are fully dry. This will help to stop the spread and growth of germs. Once dry, the parts should be stored in a dry, clean bag.

- **Soak in a solution of one part household bleach and 50 parts water for three minutes.**

Rinse all parts well. Use sterile water for the final rinse. You can make water sterile by boiling it for five minutes. This boiled water should only be used once.

Air-dry the nebulizer parts by placing them on a clean paper towel and covering them with another clean paper towel. You should replace the paper towels with dry ones when they become wet until the parts are fully dry. This will help to stop the spread and growth of germs. Once dry, the parts should be stored in a dry, clean bag.

- **Soak in 3 percent hydrogen peroxide for 30 minutes**

Rinse all parts well. Use sterile water for the final rinse. You can make water sterile by boiling it for five minutes. This boiled water should only be used once.

Air-dry the nebulizer parts by placing them on a clean paper towel and covering them with another clean paper towel. You should replace the paper towels with dry ones when they become wet until the parts are fully dry. This will help to stop the spread and growth of germs. Once dry, the parts should be stored in a dry, clean bag.

**The cups available for your medications are as follows:**

- Sidestream nebulizer cups are used for pulmozyme, albuterol and hypertonic saline.
- Pari nebulizer cups can be used for pulmozyme, albuterol, and inhaled antibiotics like Tobi and Cayston.

**These recommendations were taken from the CF Foundation website:**

<https://www.cff.org>



*CFLF grant recipient Mika enjoying a day on the river*

## Meet the Cystic Fibrosis Lifestyle Foundation

**By Erin Evans, CFLF Program Coordinator**

Many of us know about the Cystic Fibrosis Foundation, but have you heard of the Cystic Fibrosis *Lifestyle* Foundation (CFLF)? Started by a 39-year-old with CF, this group helps people live healthy and active lifestyles so they can live “stronger and longer” with CF.

What can the CFLF do for you and your family?

First, they offer Recreation Grants to people with CF who need help paying for physical activities like a gym membership, horseback riding lessons or a summer camp. So far, the CFLF has helped over 800 people pay for the activities they need to stay healthy!

To learn more about recreation grants and how to apply, visit:

<http://www.cflf.org/recreation-grants>

You can also check out the CFLF’s blog, which has new stories each week written by people with CF or affected by CF. I love these stories because they are about real life with CF, and they offer tips that are both practical and inspirational.

To read the latest blog articles, visit: <http://www.cflf.org/blog>

Interested in making a donation to the CFLF?

<http://www.cflf.org/donate-now>

## New Quality Improvement Project in Adult Clinic

**By Maryann Ludlow, RD, CDE, Clinical Dietitian**

The adult cystic fibrosis clinic has started a quality improvement project aimed at improving diabetic control in our patients with cystic fibrosis related diabetes (CFRD).

PORT CF data in 2014 revealed that adult UVM Medical Center patients with CFRD have overall poorer lung function than those UVM Medical Center patients without CFRD. They also have poorer lung function compared with CFRD patients at top-performing national CF centers.

We aim to change this situation for the better! Our goal is to improve CFRD control in all adult CFRD patients at UVM Medical Center by achieving individual annual average HbA1c of 6.5 percent or lower as of July 1, 2017.

The first phase of this project is to gather information from our CFRD patients. This will help us find out what they think some of the barriers are to having their best diabetes control. We are starting with a survey that we are distributing to CFRD patients when they come to clinic. The results of the survey will help us to focus our efforts on helping patients address the barriers to better diabetes control that are the most relevant to them.

We will keep you posted as this project progresses!

## Patient art on display

By Kelley Weston

For the past year, two third-year medical students at UVM have created body maps with children and adults with cystic fibrosis. Brett Powers and Kelley Collier worked with patients during their hospitalizations at UVM Medical Center. Body mapping is a form of artistic expression. It allows participants to share a physically or emotionally challenging aspect of their lives through a visual representation of their own body. It involves drawing (or having drawn) one's body outline onto a large surface. Then one uses colors, pictures, symbols, and words to represent experiences lived through the body. Please stop by the exhibit outside the Children's Specialty Center and check out some amazing patient artwork!



### Patient Perspective on Body Mapping

My name is Emily, and I am 17 years old. During my last hospitalization I was asked if I wanted to participate in a body mapping project. I was interested in being part of a project that could help raise awareness of CF, and I wanted to positively help others with CF, since it can be isolating to not come into close contact with other kids with CF. This was a way to be together in a different way. We started by tracing my body on paper, and I could fill it in with anything I wanted to represent myself. I chose to draw sixty five roses as my lungs and an IV bag of glitter going through my veins. I also have had a few surgeries resulting in scars, which I wrote the word *strength* on, because I like to think of my scars as my way of being stronger than something that could have hurt me. The project was a nice distraction from the normal hospital routine and can help kids who have hospital stays deal with how they feel towards their illnesses.



Artwork on display in front of the Children's Specialty Center

### CF Resource Corner (continued)

- Milestones in CF Care: Newly Diagnosed/Early Childhood book 1 of 3 - Each book is tailored to a specific developmental level. Included are quotes from CF families, transition checklists, contacts, calendars, calorie planners, therapy trackers, questions to ask the care team, teaching techniques and responsibilities, parent-teacher communication tips and advice.
- Salt Administration Handout (for children under the age of 1)
- Child Life Specialist Flyer - A document explaining the roles and responsibilities of a CLS including education, support and advocacy.



## The AFA needs you!

By Laurie Eddy

In late 2014, the Cystic Fibrosis Foundation introduced the CF Adult and Family Advisors (AFA), a group created to allow the Foundation and its partners to include more people with CF and their families in programs and initiatives that interest them. Today, there are nearly 250 members in the CF AFA group. The CF Foundation is always accepting applications and hopes to grow the diversity of the group — not only geographically, but by gender, connection to CF, ethnicity, language and experiences. If you are interested in providing your unique perspective about CF care, research and quality of life, please apply at [afasignup.cff.org](http://afasignup.cff.org).

### Insight Research Project

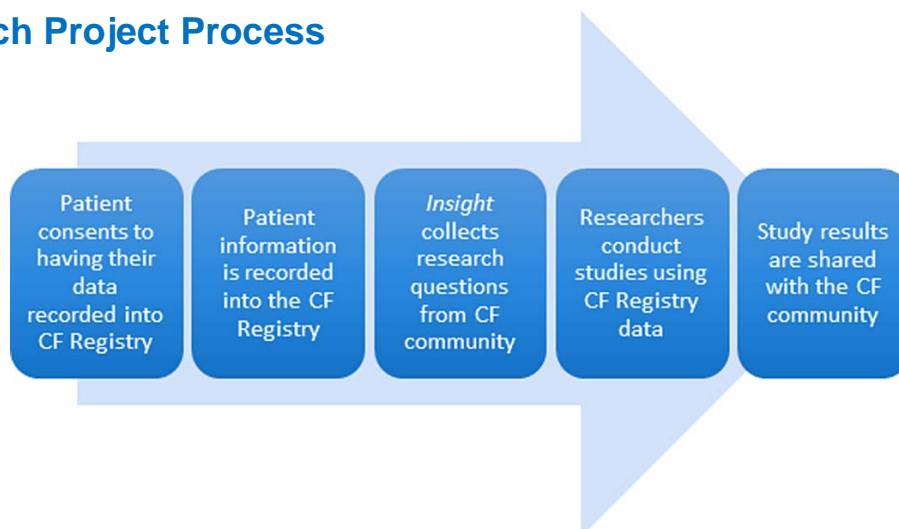
In April 2015, I signed up to be a member of the CF AFA and was invited to serve on the Patient and Family Research Advisory Committee (PFRAC). There are two things that I love about this committee, 1) the people: the group includes four people with CF, four parents, one sibling, and two spouses from around the U.S. as well as researchers from the CF Foundation, and 2) the work: we are asking the CF community to tell us what research questions are important to them.

Over the past year, this committee has been working on creating the **Insight Research Project**. Insight is designed to use the CF Patient Registry data to answer questions submitted by the CF community. The CF Patient Registry dates back to the 1960s, and the data includes the health status of over 28,000 people with CF who receive care at CFF-accredited care centers. The Registry data is already being used to help improve the care for people with CF. Each year about 15-20 new projects are started. A few examples include the impact of nutrition on children's lung function, factors that influence lung function decline in adults, and use of Pulmozyme in combination with hypertonic saline. The results of these studies are published and shared with the CF care centers to improve patient health outcomes.

Coming soon is your opportunity to submit questions. Be on the lookout for an invitation from Insight to submit your research questions in an easy online process. Questions submitted will be narrowed down to those that can be answered by data from the Registry. Researchers will then conduct studies using Patient Registry data, and results will be shared with CF community.

What are your research priorities? What areas of CF research are important to you? Contributing your voice could directly impact the future of CF research.

## Insight Research Project Process





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## News from the NNE Chapter of the Cystic Fibrosis Foundation

The Northern New England Chapter of the Cystic Fibrosis Foundation is excited to announce that as of August 1 there is a full-time development manager living in Vermont. He will work with our event committees, chairs, and sponsors.



Jim Gilbert started with the Northern New England Chapter in May of 2015. He has been working on the Annual Fund, the New England Classic Charity Trail Ride, and Great Strides.

Jim is originally from Fairfax, VT and is a graduate of both Champlain College and Johnson State College. Prior to joining the Foundation he worked for nine years at the UVM College of Medicine Development and Alumni Relations Department. We are very excited to have a dedicated staff member for Vermont and can't wait for all of you to meet Jim!