FALL 2017

THE University of Vermont MEDICAL CENTER

THE University of Vermont Children's Hospital

CF CONNECTION

Newsletter of the Vermont Cystic Fibrosis Center Advisory Board

TO MASK OR NOT TO MASK?

By Liz Hammel, VT CF Center Advisory Board

A few years ago, the staff of the Vermont CF Center began recommending that people with CF put on a surgical mask whenever they enter the hospital for clinic visits, hospitalizations, or for any appointment in a health care setting. Like any recommendation, some people follow this one and some don't. To help us decide to mask or not to mask, it might be helpful to learn more about why the change came about. Wearing a mask is an infection-prevention suggestion that came directly from the Cystic Fibrosis Foundation.

In 2013, the Cystic Fibrosis Foundation updated its guidelines for infection prevention and control. A decade of research and studies had uncovered effective strategies to prevent transmission of germs from person to person. There was also greater understanding of how respiratory germs are transmitted through droplets and the air. And there were findings on how transmission of germs can lead to worse outcomes for CF patients.

The recommendation to wear masks in health care settings is meant to reduce a person's risk of breathing in others' germs. Masks can block out germ-carrying droplets or the remains of droplets that can float in the air. We are more likely to be around sick people while at clinic or in the hospital, so wearing a mask in the public areas helps lower the chances of picking up germs from other people. It is not necessary to keep the mask on when inside a clinic exam room, the person's own hospital room, or during a

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New Faces in Clinic

Heather Coons, RN, is from Georgia, Vermont and received her degree from Curry College in Boston. She returned home to Vermont and worked in the NICU here at The University of Vermont Medical Center for five years. She is excited to step in to her new role as per diem pulmonary nurse at the Children's Specialty Center. You may also see her helping out in the cystic fibrosis clinic. She spends most of her free time with her two children, 11 months and 2-1/2 years old. They enjoy swimming and cooking together.

Lauren Elizabeth (L.E.) Faricy, M.D. is excited to join the CF team as a pediatric pulmonologist. Dr. Faricy completed her pediatric residency training at the University of Vermont and her fellowship training at the University of California San Francisco. She is particularly interested in promoting exercise and exploring the use of exercise testing in children and teens with cystic fibrosis. She and her family have moved across the country four times in the last decade and are delighted to finally land back in the Green Mountains.

To Mask or Not to Mask

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pulmonary function test.

Another recommendation from the latest guidelines is that people with CF keep a safe six-foot distance from others with CF. This is because germs can spread that far by droplets that fly through the air when someone coughs or sneezes. People with CF should also observe the six-foot rule when near anyone with a cold, flu, or infections. This is more likely to happen in a hospital setting.

A third guideline is that everyone (patients, parents, friends), in addition to clinic staff, clean their hands before entering and leaving a clinic or hospital room. We can catch and spread germs when we touch a surface with germs on it, including doorknobs, pens and handrails – and then touch our eyes, nose or mouth. Cleaning hands upon entering a clinic or hospital room helps to prevent bringing other people's germs into that space with us.

All of these guidelines are meant to protect people with CF, similar to the medications prescribed and types of airway clearance we teach. The clinic staff knows that all of these things help keep people with CF healthy and active.

No one is going to force a person to wear a mask around clinic or the hospital, just like they won't follow patients home and force them to take their meds or do their chest PT. They do their best to give people with CF and their families the best advice and leave it to us to make the choice to follow that advice.

Just like we wear seat belts to protect us from injury in car accidents, condoms to protect us from sexually transmitted diseases, and sunscreen to protect us from damaging UV rays, masks protect people with CF from other people's germs. It is smart to wear one while at clinic and the hospital.

Time to Get Your Flu Vaccination!

The influenza season is approaching again. It is very important that all patients with CF, as well as their family members or others in their home, receive the influenza vaccine. Within the next few weeks, the hospital will receive its supply of flu vaccines. We encourage patients to receive this vaccine from their primary care physician, local flu shot clinic, or from our CF clinic.

Please don't delay. People with CF are at increased risk for respiratory complications from influenza infection, including pneumonia. The influenza vaccine helps to reduce the risk of these complications.

CF SCHOLARSHIP FOR 2017

The University of Vermont Medical Center is pleased to announce that Emily Woodward was awarded the 2017 CF Scholarship. Emily will attend Colby-Sawyer College in the fall and will major in nursing.

Emily wants to specialize in pediatric care. She was inspired by the high quality of care that she has received at UVM Children's Hospital and our CF clinics. Congratulations, Emily!

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 available age-appropriate educational materials and generally helpful resources. Watch as the list grows over time.

The names of the newest resources are in **bold**. How many of the below resources do you have? How many have you browsed or used recently? Please feel free to reach out if you are missing one or more of the below resources. They are meant to be tools for caregivers, siblings, teachers, nurses, etc. We can provide extra copies if needed.

- 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
- MyHealth Online brochure
- CFLifeLessons.com video clip
- ***Living with CF Series*:** Eight pamphlets created by CysticLife and Parenting Children w/Health Issues. Included are helpful tools, perspectives, ages and stages, do's and don'ts, health maintenance recommendations, school advice, transition information, advice from others, and additional resources. Written for parents of infants through parents of teens.
- ***Huxi: A Coloring Adventure*:** The amazing stories of Huxi, a panda with CF, are educational resources that explain how CF is obtained, the daily regimen that is necessary to manage CF and prevent illness, as well as the importance of maintaining a full and active life. Written for preschool age to early school age.
- ***An Adult Coloring Activity*:** An interactive book that includes inspirational quotes and coloring pages geared toward older teens and adults.

Understanding the Oral Glucose Tolerance Test and CF-Related Diabetes

By Paul (P.J.) Zimakas, MD

Cystic fibrosis-related diabetes (CFRD) is an important and common complication of cystic fibrosis. CFRD generally occurs in individuals 10 years of age or older. The frequency increases dramatically with age. In fact, CFRD is now the most common non-respiratory complication of CF encountered among individuals school-aged and older.

Diabetes is a condition that interferes with how your body is able to use sugar for energy. Every cell in your body requires energy to work properly. This energy is needed to keep your brain thinking, your heart beating, your lungs breathing, and your muscles working. The preferred source of energy for the body is the most basic form of sugar, called *glucose*. When you eat food, much of it is digested and absorbed into the bloodstream as glucose. The body can even make its own glucose from an organ called the liver.

The bloodstream transports glucose to the different parts of the body. But, in order for glucose to enter the cell to be used for energy, you need to have a hormone called insulin. Insulin is like a key that opens the door to your cells so that the body can use whatever glucose it needs for energy. Your body then stores any extra glucose that it does not need right away for times when you are not eating food (fasting).

Diabetes results when insulin cannot do its job properly. As a result, without the effect of insulin, the body cannot use glucose. It just accumulates in the bloodstream, causing “high blood sugar.” Cystic fibrosis causes diabetes by damaging the cells in the pancreas that produce insulin. CF pulmonary infections also cause resistance to insulin. As a result, in CFRD we see both low levels of insulin and an inability to respond to that insulin properly.

Some, but not all, people who developed CFRD may begin to notice symptoms or signs. These include having to urinate frequently, feeling extremely thirsty, losing weight or difficulty gaining weight appropriately, or having unexplained decline in their pulmonary function tests. However, some individuals may have no obvious symptoms or signs in the early stages of CFRD. The reason why your CF team is so concerned about the development of CFRD is that it is associated with worse lung function, difficulty maintaining a good weight, more chest infections, and decreased survival.

Even those who have developed diabetes, but do not have any symptoms, may start to suffer from their CFRD with respect to poor lung function and long-term health. Early detection and treatment can improve greatly the long-term outcome for these patients. Therefore, all individuals with CF who are 10 years of age or older should be screened for diabetes once per year. We use an oral glucose tolerance test (OGTT) to do this.

The OGTT consists of measuring the fasting blood-sugar level (referred to as blood glucose level) in the morning and then drinking a fixed amount of sugar. This is usually in the form of orange or cola syrup provided by the testing lab. The blood glucose is then repeated two hours later to complete the test. For some who are unable to tolerate the taste or consistency of the liquid, your CF team has a few other options to choose from.

The results of the OGTT will generally fall into one of three categories:

1. Normal blood glucose handling: No evidence of CFRD.
2. "Pre" diabetes: Also known as "Impaired Glucose Tolerance.” These results indicate that the pancreas is straining to produce enough insulin to control the blood glucose levels. It is often a warning sign that diabetes may soon develop.
3. CFRD: The blood glucose levels are significantly elevated and fall within the range considered diabetic.

A single, abnormal OGTT may not indicate the need for treatment, but it can identify individuals with abnormal glucose tolerance and the need for close monitoring. Close monitoring may include repeating the OGTT within a year, frequent monitoring of blood glucose during pulmonary infections, and attention to changes in nutritional or pulmonary status.

If results of testing are abnormal, a consultation with an endocrinologist (a diabetes specialist) will likely be requested. At

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a minimum, starting to test the blood glucose levels at home on a regular basis will be needed. Caregivers or older children will be taught how to perform this test on a drop of blood obtained from the fingertip. The blood glucose level is typically checked prior to eating breakfast and two hours after the largest meal of the day. Frequency of testing can range from two-to-three times per week to multiple tests on a daily basis.

Individuals with abnormal results on their OGTT will also be counseled regarding some dietary changes. This means avoiding foods and beverages that are known to cause spikes in blood glucose levels and serve as an extra stress on the already strained pancreas. Individuals with any degree of abnormal glucose tolerance are ideally managed with a diet that provides necessary calories for maintaining appropriate weight but limits the amount of simple or rapid-acting sugars. Most notably, this includes avoiding high-sugar beverages.

If there is conclusive evidence that diabetes has developed, then treatment will begin. The only proven therapy for CFRD is insulin injections, often given multiple times per day. Unfortunately, there is no effective pill or inhaled form of treatment for diabetes. As part of your CF team, your endocrinologist and diabetes nurses will serve to educate, inform and help manage the CFRD.

There is no denying that the treatment of CFRD adds substantially to the daily burden of managing cystic fibrosis. Rest assured that your entire CF team is very sensitive to this fact and will be there to support you in managing this. Like all of your other recommended CF treatments, the treatment of CFRD definitively has beneficial effects on nutrition and improves pulmonary function and survival.

The Impact of Treatments and Quarterly Visits on Funding

By Tom Lahiri, MD

We in the CF community are living in an exciting time! There are new drugs (Kalydeco and Orkambi) that are able to modify CF disease in a large number of patients. Over the next few years, we expect that new, possibly better, drug combinations will be available to most of our CF patients. Many of you have asked whether your (or your child's) other chronic treatments, such as Pulmozyme, hypertonic saline, ibuprofen, azithromycin and even airway clearance, can be eliminated or substituted when you (or your child) start a CFTR modulator therapy like Kalydeco or Orkambi. The simple answer is that we just don't know.

At this time, we are not recommending discontinuation of any recommended treatments or medications, as the long-term effects of the newer drugs is unknown. We will continue to offer patients all of these other therapies even if they are on Kalydeco or Orkambi. It is also critical that you come to your scheduled quarterly appointments so that any side effects of the newer therapies can be monitored. Coming to visits is the best way to be proactive about your health. We are expected to follow strict criteria to maintain accreditation by the Cystic Fibrosis Foundation. All patients in the CF Patient Registry are expected to be seen four times per year and have certain monitoring tests performed. Our funding from the CF Foundation is directly linked to how well we follow these guidelines.

If you have any questions about why you or your child has been prescribed a certain treatment or therapy, please ask of the CF providers. We are here to help!

The *Insight CF* Project

By Laurie Eddy, VT CF Center Advisory Board

Late last year, the CF Foundation's Patient and Family Research Advisory Committee launched the *Insight CF* Registry Research Project. It is the CF Foundation's first community-driven research initiative. Everyone in the CF Community was invited to get involved and add their voice to the work of the CF Foundation.

We began the *Insight CF* project by asking people in the CF community to submit research questions that could be answered through the CF Foundation Patient Registry. More than 350 people submitted questions, and more than 400 questions were asked! The questions were sorted into three categories:

1. Questions we already know the answers to
2. Questions that cannot be answered by the registry (they would require more research)
3. Questions that can be (but have not already been) answered with data already in the registry

All submitted questions will be used to help us understand the community's needs and to help inform the research moving forward. The questions that fell into the first category will be shared across the CF Foundation to assist with content development on the foundation's website. The questions in the second category – those that would require more research outside of the registry – will be forwarded to researchers for consideration.

The questions that fell into the third category came back to our *Insight CF* project team. Adults with CF, parents, siblings, and spouses made up this team. We worked to condense those 150 questions into a list of 22 questions.

In June, we distributed a survey asking members of the CF community to vote on which question was most important to them so that we can answer the top questions by using the registry. The survey received 1795 responses, which was the highest response received of all CFF surveys.

Who responded to the survey?

18% (309 responses) were people with CF

38% (687 responses) were parents (32% were mothers, 7% were fathers)

18% (301 responses) were relatives

25% (420 responses) were professionals

What questions were rated in the top three?

1. What are the factors that predict long-term survival?
2. Geography: How do geographic and environmental factors such as climate, altitude, and air quality impact lung infections and health outcomes?
3. Lung function: What are the predictors of lung function decline, and do the predictors change as people age?

What happens next?

A multidisciplinary research team will develop a study design and analysis plan for the chosen research questions. The plans will be submitted to an Institutional Review Board to ensure the protection of the rights and wellbeing of participants. Once approved, the research team will conduct the analyses of data from the registry. Results of the research will be disseminated to the community through CFF.org, and will also be submitted for publication in scientific journals.

Insight CF *will be at the NACFC!*

The Patient & Family Research Advisory Council will present a poster at the North American Cystic Fibrosis Conference in November to share the *Insight CF* Project with researchers and health care professionals from around the world.

2016 CF Registry Data Report

By Tom Lahiri, MD and Laurie Leclair, MD

Each year, the Cystic Fibrosis Foundation reports on how CF centers across the United States have performed in the areas of nutrition, lung function, adherence to CFF guidelines, and prevention. The Vermont Cystic Fibrosis Center received its latest registry data from the CFF earlier this summer. Dr. Lahiri and Dr. Leclair have summarized information about our pediatric and adult programs, respectively.

Pediatric CF Registry Data Report

Nutrition

The pediatric program's median BMI percentile (body mass index) has gradually improved to the 64.2 percentile. This is approaching the top 10 centers nationally.

The nutritional status (weight for length) for infants and children less than two years old improved for the third consecutive year. It is now above the national average.

The nutritional health for children with CF-related diabetes (CFRD) remains among the highest in the U.S.

Pulmonary

Lung function (FEV1) for children 6-17 years was stable from last year at 98.3% predicted.

Adolescent (13-17 year old) lung function remains in the top 10 centers nationally.

Lung function for children with CFRD remains in the top 10 nationally.

Our center tends to hospitalize more children for pulmonary exacerbations than the national average.

Our rates of bacterial infections (Pseudomonas, MRSA, *B.cepacia*) are stable and close to the national average.

Guidelines and Prevention

100% of children under 6 years old met screening guidelines (great job!).

Only 81% of 7- to 17-year olds met guidelines (seen four times with PFTs, cultures). This is a further decrease from last year and is our lowest level in ten years.

Screening for CFRD has increased but we are still missing children (78% screened).

About 20% of our children live in a household where at least one parent is a smoker.

Adult CF Registry Report

Nutrition

The adult program's median BMI remains very near the national average at 22.8. It increased from 22.4 last year.

The median BMI for patients without CFRD is higher than those with CFRD (23.4 vs. 21.6). It emphasizes the need for ongoing additional support for our patient with CFRD.

A dietitian saw 100% of the adult patients who are below their nutritional goals. This reflects our increased dietitian support for the adult program.

Pulmonary

The adult program's median FEV1 from 18-29 years old is 81.5%. This is nearly 10% better than the national median.

Median FEV1 for adults older than 30 is 65.5 and remains close to the ten best centers in the registry.

Lung function in CFRD patients remains lower than our non-CFRD patients. Our adult program's median, however, has improved to be near the national median.

Guidelines and Prevention

We have seen a steady increase in percent of patients being evaluated by all members of the CF team. This likely reflects our improved staffing. Only 72.4% of adult patients met the CF guidelines of four visits, at least one culture and two PFTs. This is a significant improvement from last year (up almost 20%). Great job getting to clinic!

We are near the top performing centers in oral glucose tolerance test screening for CFRD. Nice job!

Copay Assistance for CF Medications

Do you have high copays or do you want to save a little extra money each month? Here is a list of copay assistance programs available for CF patients. Please contact Christine Prior, LICSW Christine.prior@uvmhealth.org with any questions or assistance.

Creon Care Forward: www.creon.com/cfcareforward

- Copay: $0 or $5 (if over $75)
- One free MVI (MVW Complete or Libertas) and one nutritional supplement per month
- Must have commercial health insurance; not eligible under Medicare, Medicaid, or Tricare

Zenpep Live 2 Thrive: www.live2thrive.org

- Copay: $0, if < 2 years old
- Copay: $0 – $40 (if cost is $500, copay = $40)
- One free MVI (MVW Complete), one D3 vitamin, and one nutritional supplement per month
- Must have commercial health insurance; not eligible under Medicare, Medicaid, or Tricare

Pertzye: Chiesi Care Direct: https://www.pertzyecf.com/savings-support/

- Copay: $0 - $20
- $75 debit card provided with each month's fill, up to $900 per year
- Must have commercial health insurance; not eligible under Medicare, Medicaid, or Tricare

Cayston Access Program: https://www.cayston.com/cayston-access-program/

- Co-pay: $10
- Must have commercial insurance; not eligible under Medicare, Medicaid, or Tricare

TOBI Podcare+: http://www.tobipodhaler.com/info/about/podcare-cf-patient-support.jsp

- Copay: $0 Podhaler caps
- Copay: $4 TOBI solution
- Must have commercial insurance; not eligible under Medicare, Medicaid, or Tricare

Bethkis: Chiesi Care Direct: https://bethkis.com/savings-support/

- Co-pay: Eligible patients pay as little as $0
- $50 debit card provided with each month's fill
- Must have commercial health insurance; not eligible under Medicare, Medicaid, or Tricare

Pulmozyme Access Solutions: http://www.pulmozyme.com/pulmozyme-copay-patient-financial-support

- Co-pay: $30 or less
- Must have commercial insurance; not eligible for Medicare, Medicaid, or Tricare

Kalydeco/Orkambi: Vertex GPS: https://www.vertexgps.com/

- Co-pay: As little as $15
- Must have commercial insurance; not eligible for Medicare, Medicaid, or Tricare

Health Well Foundation: https://www.healthwellfoundation.org/

- Must have insurance; open for commercial, federal, or state funded insurances
- Two grants: Treatments and vitamins and nutritional supplements
- Used in conjunction with other copay programs to bring copay to $0 for CF medications
- Pays for vitamins and nutritional supplements
- Income limit of 500% above the federal poverty line

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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.

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 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

You can change the future of cystic fibrosis treatment. Find an enrolling clinical trial with the Clinical Trial Finder. https://www.cff.org/Trials/finder

Want to help plan cystic fibrosis fundraisers in your community? Contact your local chapter to get involved. no-new-eng@cff.org