

Inspire

Spring/Summer 2013

A newsletter for the Rochester Cystic Fibrosis Community

Proposed Infection Control Guidelines

Clement Ren, MD

One of the fundamental problems in CF is that infection settles into the lung tubes. In most cases, the bacteria that are responsible for these infections comes from the environment, but we also know that infections can potentially be passed from one CF patient to another. To help protect the health of CF patients, the CFF published infection control guidelines in 2003. In the 10 years that have passed since those guidelines were published, new information has become available, and this has led to a new set of guidelines being developed. These guidelines address 3 key areas for infection control: public events, the outpatient clinic, and the inpatient hospital environment. The draft guidelines were recently released in March, and the CFF will be opening a period of public comment in May. The guidelines will be finalized this fall. However, in the meantime the CFF has already released recommendations for public events. These are available on the CFF website at <http://www.cff.org/aboutCFFoundation/InfectionPreventionControlPolicy/Policy/> and are summarized here:

- ◆ Only one person with CF may be present at a CF Foundation-sponsored indoor event, meeting or office.
- ◆ At CF Foundation-sponsored outdoor events or gatherings, people with CF need to maintain a distance of at least 6 feet from each other.
- ◆ Under no circumstances shall individuals who have ever had a confirmed positive sputum culture for *Burkholderia cepacia* (B. cepacia) complex attend any CF Foundation events, meetings or offices.
- ◆ Everyone (people with CF and those who do not have CF) should clean their hands using soap and water or an alcohol-based hand gel that is at least 60% alcohol. If hands are visibly dirty, use soap and water. Hands should be cleaned after coughing, sneezing or blowing the nose, before eating, after going to the bathroom.
- ◆ Everyone should cough/sneeze into a tissue, immediately discard it in a no-touch trash/waste receptacle and immediately clean their hands.
- ◆ All people with CF should avoid the following activities and risk factors with others with CF because of the risk of transmission of CF germs in non-health care settings. These include, but are not limited to:
 - Shaking hands, hugging or physical contact
 - Kissing or intimate contact
 - Riding in the same vehicle or sharing a hotel room
 - Fitness/exercise class at the same time
 - Sharing personal items, drinking cups, utensils or respiratory equipment
- ◆ Information about whether a person has CF, or what germs might be in the individual's lungs, is to be maintained as confidential medical information unless the family chooses to make this information known.
- ◆ The CF community should have education regarding infection prevention and control for CF. Materials are available on the CF Foundation's website and through CF Foundation-accredited care centers.

These are very significant changes to the previous policy, and they were made by the CFF based on the new data available and the goal of protecting and maintaining the health of all CF patients. Further information about the new guidelines and the reasons behind the revision can be found at the CFF website, www.cff.org.

Here in Rochester, our CF team is planning to meet in June after release of the guidelines to make a plan for how to implement these changes in our public events, outpatient clinic, and hospitalized patients. We plan to work with patients, families, nursing, and the URMIC infection control staff to implement these changes in a way that meets the goals of infection control without being excessively onerous. Just like the CFF, our goal is to protect and maintain the health of all CF patients while optimizing their quality of life. Please do not hesitate to contact us if you have any questions or concerns about these new guidelines.

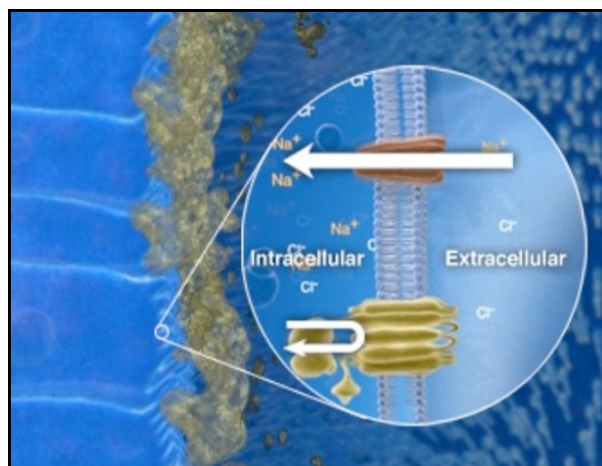
With the exciting new advancements in CF medications, especially Kalydeco, and the new ones that are currently in clinical trials, we want our families to know how CFTR (CF transmembrane conductance regulator) potentiators and correctors work and why these medications are currently only available to a small portion of people with CF. These are the first medications that target the basic defect in CF and are in pill form! There are about 1800 known CF genetic mutations, and each is unique. CF and genetics are very complex and the CF community is fortunate to have brilliant scientists who make these discoveries!

In the airway, chloride ions are secreted through CFTR channels. CFTR works in the lungs, pancreas, GI tract, reproductive system, liver, and kidney. Sodium and chloride are balanced in the body thanks to the help of these channels working together. The balance of sodium (Na) and chloride allows the surface of the airway (airway surface layer) to have enough fluid to work well and help the cilia on the lungs move freely. Cilia are the tiny hairs in the trachea and parts of the lungs that help clear germs and tiny particles and move them up toward the mouth, in order to maintain a healthy airway.

CFTR genetic mutations that cause CF can affect the:

1. Quantity of CFTR channels at the cell surface
2. Function of CFTR channels at the cell surface

In the sweat glands, dysfunctional CFTR restricts reabsorption of chloride limiting the amount of sodium and chloride that can be reabsorbed. The result is a high concentration of salt in the sweat. CF genetic mutations (example: $\Delta F508$) that affect the quantity of CFTR channels at the cell surface can lead to complete lack of channels or lead to a reduction in the number of channels present at the cell surface. CF mutations that reduce CFTR channel function may result in complete loss of function or a reduction in the function for channels that are at the cell surface of the airways. Mutations affecting quantity can range from those that cause little or no CFTR channels reaching the cell surface to few channels reaching the cell surface.



Regardless of the problem, CFTR dysfunction leads to similar results. There is a lack of balance of chloride and sodium which results in the dehydration of the surface of the cells and in a decrease in the ability of cilia to work to clear the area of debris. Mucus gets thicker and stickier due to the dehydration of the airway surface layer. This leads to an increase in viruses, bacteria, particles, and air trapping in the lung, which causes infection and leads to lung damage.

It is by targeting these defects that has led to the development of CFTR potentiators, Kalydeco (aka ivacaftor) and correctors (aka lumacaftor). Kalydeco alone works only in those with the G551D CF mutation, as there is a gating defect in the CFTR protein. This means that there are normal quantities of CFTR channels, but they are unable to open. The administration of Kalydeco allows for the opening of these channels so that adequate chloride can get to the surface of the cell. G551D only occurs in 4% of people with CF.

CFTR mutations that affect conductance result in a normal quantity of channels at the cell surface; however, the ability of chloride ions to flow through the channel is reduced. R347P is the most common example of a CF mutation that reduces the rate of Cl flow through CFTR. There is still partial chloride function.

So, as you can see, depending on the CF mutation, there are difficulties with chloride ion transportation. Fortunately, these drugs are in the process of being further developed in clinical trials, including one at our center, to determine their safety and efficacy. For more information, go to CFF.org and check out the research pipeline and more on the Vertex clinical trials.

You have probably noticed that your CF providers have been discussing testing for diabetes more frequently over the last few years. This is because there are guidelines from the CF Foundation recommending regular testing for diabetes in all patients with CF. Hopefully after you have read this article you'll have a better understanding about CF-related diabetes, why your provider is recommending screening, and why your provider may be discussing treatment.

There are two common types of diabetes, type I and type II. Type I diabetes is a condition where a patient is unable to make enough insulin to control their blood sugars and Type 2 diabetes is a condition where a patient has enough insulin but it doesn't work well because of resistance (usually this is because of being overweight). CF-related diabetes is closer to type I diabetes since patients with CF have some scarring of the pancreas over time and they don't make enough insulin to help control blood sugars. In many cases however patients with CF have some insulin resistance because of lung infections or treatment with steroid medication during lung infections.

Many doctors who take care of patients with CF believe that having high blood sugars can affect lung function and lung infections and can interfere with the ability for patients with CF to gain weight. There are some studies that suggest the CF patients who have diabetes and don't control their blood sugars have more infections and hospitalizations and lower lung function over time than patients with CF who don't have diabetes.

CF-related diabetes can start in adolescence or young adulthood and can be seen in the anywhere from 20-50% of patients with CF. Many patients with CF-related diabetes have no symptoms at all and the high blood sugar is found when routine tests are ordered at your clinic visits. The most common symptom that patients with diabetes have are excessive thirst or excessive urination but these symptoms are often not present. Since symptoms are often not present the CF foundation is recommending screening (testing) for diabetes every year starting at 10 years of age.

CFRD Screening Guidelines

- Annual 2 hour OGTT beginning at age 10 during baseline health.
- During acute illness (tune-ups) - fasting and 2 hour post meal glucose checked for the first 48 hours when inpatient.
- For those on overnight tube feeding —blood glucose checked mid feed and immediately at the end of feeding at least once per month.
- 2 hour OGTT when planning pregnancy or when pregnancy confirmed. OGTT again at 12-16 weeks AND 24-28 weeks.
- 2 hour OGTT within 6 months of lung transplant

Your provider will talk with you about the tests that are available to test for diabetes, there are simple blood tests, finger stick tests and a glucose tolerance test that may or may not require you to fast before the test. The tests are all basically looking for a higher amount of glucose in your blood. After a thorough study of the different tests the CF foundation has decided to recommend a test called the "2 hour oral glucose tolerance test (2 hr OGTT)". This test requires an appointment in the lab so they can have the glucose solution that you must drink for the test. On the morning of the test you arrive at the lab on an empty stomach and they draw your blood, after that you drink all of the liquid and you wait 2 hours to have your blood drawn again. Sometimes these tests will provide a "warning" to your provider that you are at risk for diabetes and sometimes your provider will conclude that the test means you have diabetes. In either of these cases it will lead to an open discussion with you about what that means for your health and if any treatment is needed.

Since good nutrition in patients with CF is so important you will no doubt talk with your provider and with your CF dietitian about what having diabetes and CF means. You probably know that for patients with diabetes who don't have CF there are lots of restrictions about what should/should not be eaten, this is not the case for patients with CF-related diabetes. For patients with CF we continue to recommend eating a healthy CF diet and often recommend extra calories, while you may have discussions about how to know how many carbohydrates is in your diet it is more for helping your provider and dietitian decide on treatment. Often the first step in deciding about treatment is getting more information from you about your eating habits. Frequently we will be discussing the idea of checking your blood sugars at home by finger stick to help us understand at what part of the day you are having the highest blood sugars.

Continued next page

CFRD continued

If you and your providers decide on treatment with medications this is almost always treatment with injections of insulin in patients with CF-related diabetes. While oral medications do work for patients with Type 2 diabetes they aren't used in CF-related diabetes. There are two main types of insulin that are used and the number of injections per day that are recommended depends on your blood sugars and how much treatment you are willing to consider.

Your Stories: Caitlyn Casey

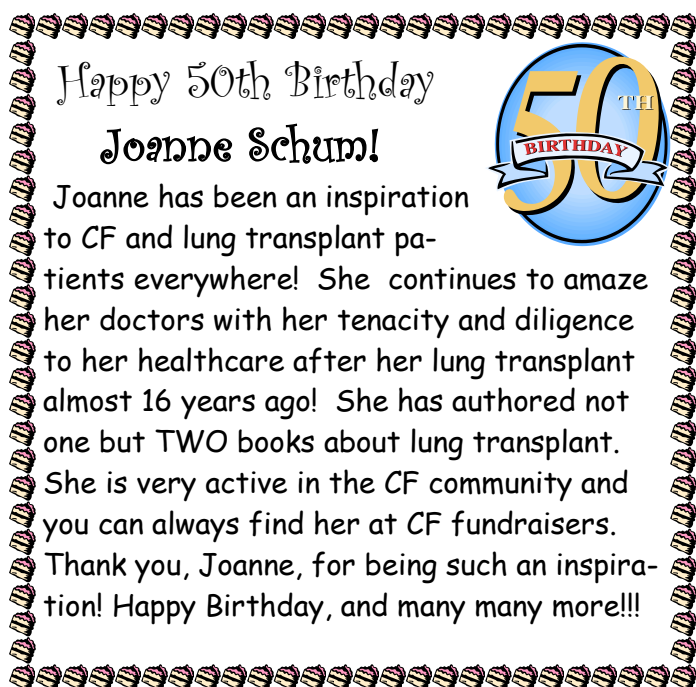
Having CF is hard work. Besides all of our regular treatments and medications, different therapies are often added as we get older. When I first learned I had developed cystic fibrosis related diabetes (CFRD), I was annoyed and didn't want to be bothered with another medication or think I could fit another routine into my schedule. Plus, I had feared diabetes since being a child because of the shots and needles! But I didn't have a choice- I had to accept the diagnosis or lose my health. As I continue my walk with CFRD, I'm learning it is nothing to fear and I actually (gasp!) ENJOY some of the aspects of it...

The first pleasant surprise associated with my CFRD was the cost of supplies and equipment. I received a huge box of supplies and my total due was under a TOBI co-payment. The cost of CFRD medication is manageable and a lot less than I pay for any CF medication for my lungs. This was a huge relief for me since I pay so much already for medication.

Another pleasant surprise I encountered after starting insulin was the LACK of pain when checking my sugars and giving myself shots! In fact, the first shot I administered, I thought I did something wrong because I didn't even feel it go in. I thought, "Did I put the needle on wrong and it went back into the casing somehow?" But when I pulled the shot back, the needle was indeed leaving my belly. No discomfort, no pain! That is still the case after a few months of being on insulin- I think I have had only a shot or two that have been uncomfortable.

The best thing about my CFRD diagnosis and treatment has been the sense of control I feel I regained over my body. Having control over one's health is often rare to a CF patient. Our bodies are unruly, stubborn and headstrong but having CFRD actually gives you the ability to directly manipulate your sugars and will therefore greatly influence your CF health! Conversely, one's "numbers" might also reveal when we are getting sick. Monitoring CFRD gives you better insight into how your body is working.

Don't fear CFRD- we CF'ers can handle anything!



Congratulations Graduates!

Congratulations to our 9 patients who are graduating high school next month! Best wishes on your future endeavors!

Congratulations to our two adult patient on earning your Masters Degree!

Congratulations to our adult patient on earning your Bachelors Degree!

Congratulations to our two adult patients on earning your Associates Degree!

Such a taboo topic sometimes. Many patients (and providers!) get embarrassed and red-faced when we talk about it. But many want to know - how does CF affect sexuality and sexual relationships?



To start off, many seem to notice that development is slower than their friends. It's important to realize that everyone grows at their own pace (regardless of CF!), but we do know that puberty begins about 18 months later in those with CF than those without CF. You can help your development by making sure you are getting enough calories and nutrients.

Many have asked when the CF diagnosis should be discussed with a boyfriend or girlfriend. Only YOU can decide when it is time to tell someone about your health. You may not feel comfortable confiding in someone right away, but as the relationship deepens it may feel "right". Trusting the individual with such a personal issue may be hard for some. There are no guarantees in relationships and there may be some that feel the news of your CF is too much for them to handle. He or she may love you very much but be frightened about losing you one day.

Cystic fibrosis does not diminish sexual desire or the need to be intimate. Only YOU can decide when to become sexually active. If you decide to become sexually active, PLEASE talk to your health care provider about protection from pregnancy and STD's. For women with CF, some forms of birth control may interact with your CF medications and may not work as well to protect you from getting pregnant. Your CF provider can help guide you to making the best birth control choice for you. Other things to consider when thinking about an intimate relationship is your daily health. You may notice that you cough more doing sexual activities- it is a physical act and may cause increased coughing. Assure your partner before it happens that you are okay but may need breaks. When you aren't feeling well, you might feel more tired and your sexual energy might be low. Having a compassionate and trustworthy partner is key to any relationship and may be even more crucial with CF. Many patients have remarked not feeling "attractive or sexual" with oxygen masks and ports - this is not an uncommon feeling. These may seem like barriers to intimacy, but in a loving relationship, talking with your partner when these issues and feelings come up will help sort out these concerns. Many couples opt for counseling to help them through tough conversations.



If you want to have children or are planning to get pregnant, talk to your doctor and health care team! Pregnancy requires specialized medical attention particularly because medications may need to be changed or stopped so as not to interfere with the healthy development of the fetus. Lung function and nutrition need to be carefully monitored as well. Sexuality and CF- a taboo topic for some, but please know your health care team is here to talk and guide you through all the areas of your life which includes your sexual health.

Congratulations to our 3 adult patients who were married since our last newsletter!
Congratulations to our 2 adult patients who got engaged!
Best wishes for a happy and healthy future together!



Congratulations to our 3 adult patients on the birth of your babies!



Also congratulations to our own Mellanie Pilato-Lopez and Bridget Platania on the birth of your girls!

As we head into the summer months, many of us will be planning to take a summer trip. Whether it is a day trip to the lake or a week at the beach, the key to any successful trip is planning. Planning before you or your child's next trip to the CF clinic can also make the visit a successful one.

In December 2012, the American Thoracic Society held its second annual "Cystic Fibrosis Week at the ATS". Michael P. Boyle MD, FCCP, Director of the Johns Hopkins Adult Cystic Fibrosis Program presented a Webinar entitled, Providing Best Care for Individuals with Cystic Fibrosis: A Partnership with Patients and Families. The target audience for the webinar was patients, families and health care professionals. The three major themes presented were:

1. Does what we do make a difference or is it all genetics?
2. Partnering with your caregivers to assure you received the best possible care.
3. Strategies to help young people with CF become successful adults with CF.

One of the elements suggested as a way to partner with your CF team to assure that you or your child receives the best care possible is planning ahead for your visit to the CF clinic. A few ways to prepare for the next clinic visit to help maximize your clinic visit include:

- ⇒ Prepare a list of key questions or issues to address at the visit with the team
- ⇒ Bring ideas about improving your care that you may have heard about or have been thinking about
- ⇒ Discuss with your child what they think should be discussed at the clinic visit
- ⇒ Know and understand the numbers: FEV1 trends, weight/BMI, key labs and recent radiology reports. A "Patient Summary Report" is available at your clinic; please make sure you receive a copy at your clinic visit to take home with you
- ⇒ Ask for guidance or help with obstacles to adherence (financial stressors, life events, anxiety, depression, etc.). The CF team is here to partner with you to help you care for yourself or your child.

Speaking of planning for your next trip to clinic, please allow some additional travel time around the Elmwood/Mt Hope area when coming to Strong. There are multiple construction projects in and around the hospital, including the building of the new Childrens' Hospital!

For more information about Cystic Fibrosis Week at the American Thoracic Society, including additional links to patient care information, please visit the ATS website: <http://patients.thoracic.org/lung-disease-week/cystic-fibrosis-week-2012/>

The link to the webinar is: <http://patients.thoracic.org/lung-disease-week/cystic-fibrosis-week-2012/webinar.php>



The CF Team and our families at Great Strides on 5/19/13 at the Genesee Valley Park in Rochester. Pictured from the left: Zack Odell, Patrick Berry, Kristin Berry, Melissa Berry, Marcy Odell (& children), Karen Voter, Mary Platt, Donna Germuga, Lila Platt

Acid Suppression and the Cystic Fibrosis Diet

Rivkah Mantel Pharm.D. Candidate and Angela Nagel Pharm.D.

An important part of Cystic Fibrosis management is maintaining a balanced diet and adequate pancreatic enzyme supplementation. The Cystic Fibrosis Diet is designed to ensure proper nutritional intake. It usually consists of high caloric intake, with vitamin and mineral supplementation. In the presence of stomach acid, pancreatic enzyme supplements can be degraded. Classes of pharmacologic therapies, such as Proton Pump Inhibitors (PPIs) and Histamine 2 Receptor Antagonists (H2RAs), are prescribed to enhance the action, and prevent destruction, of pancreatic enzymes.

PPIs include Dexilant, Nexium, Prevacid, Prilosec (omeprazole), Protonix, and Aciphex; generics are available. It is important to take these medications thirty minutes to an hour prior to a meal. They should be taken at the same time each day, generally before the morning meal. Capsules may be opened and contents sprinkled over food, such as apple sauce. The contents should not be chewed or crushed. It will take several days before these medications reach their full benefit. There are a few cautions associated with PPIs such as: vitamin B12 deficiency, drug-interactions, and the possible link to gastrointestinal (GI) infection. A patient that develops signs of gastroenteritis/ moderate- severe diarrhea should contact their provider.

H2RA include Zantac, Pepcid (famotidine), Axid, and Tagamet; generics are available. These medications can be taken with or without food. If these are also being used for heartburn, H2RAs can be taken 15-60 minutes before foods known to cause heartburn, up to 2 times a day. There are chewable dosage forms available for some of these. Some precautions associated with H2RAs are: vitamin B12 deficiency, confusion (associated with kidney impairment).

Although these medications are helpfully, there are some concerns to be aware of. The most common side effects with both PPIs and H2RAs are: headache, constipation or diarrhea. Dizziness or drowsiness may occur with the H2RAs. These medications can interact with other pharmacologic therapy in a typical cystic fibrosis regimen. Be sure to consult your pharmacist whenever there are changes to your therapy. Pharmacists are a great resource for drug information and will help ensure you get the best benefit from your medications.



Vitamin Update

As many of you are aware, the manufacturing of Source CF vitamins has been suspended indefinitely. Aptalis Pharma owns the vitamin line but contracts to an outside manufacturer for production. Aptalis is looking for another manufacturer for the Source CF line but cannot predict when manufacturing will resume. For now, your CF Center is recommending switching to AquADEK vitamins. These come in gelcaps, chewable and liquid form. While AquADEK is formulated for the unique needs of a person with Cystic Fibrosis, they do not contain the same amounts of vitamins as the Source CF. This means you may need to take more of the AquADEK to get the same amount of vitamins as you were getting in the Source CF. Please contact the center for recommended dosing.

Some insurances may not cover the AquADEK. You will need to call your insurance company to verify coverage. If your insurance does not provide coverage, you can also look into whether a flexible spending account can cover the cost. If neither of these are helpful to you, please call the center for more options.

MyChart. Are You Signed Up?

What is MyChart?

MyChart offers patients personalized and secure online access to portions of your outpatient medical records; at this time, it does not include any information from hospital stays other than your patient After Visit Summary. MyChart enables you to securely use the Internet to help manage and receive information about your health. With MyChart, you can:

- View your outpatient diagnoses, medications, immunizations, and most lab and test results.
- Easily submit corrections/additions to your medications and allergies; once approved by your provider, these updates automatically appear in your URM C electronic chart.
- Request appointments and prescription renewals.
- Securely and confidentially communicate with your health care team.
- Grant access to MyChart to other adults via proxy access, as well as export portions of your chart to bring to other healthcare institutions.

How do I sign up?

To sign up for MyChart, you will need an activation code. This code allows you to log on to create your MyChart user ID and password, which you will then use to login to MyChart.

All patients over age 18 will receive an activation code at the end of each URM C visit, so you can sign up at home when it's convenient for you. You can also request an activation code online, or by contacting our MyChart Customer Service Center 8 a.m. to 5 p.m. weekdays: 585-275-URMC (8762) or 1-888-661-6162.

Patients under 18 (or their parent/legal guardian) must speak with their doctor in person before signing up for MyChart.

What type of computer do I need to use MyChart?

You can access MyChart from any computer connected to the Internet and an up-to-date browser (such as Internet Explorer). You can also view MyChart from your iPhone or Droid phones. Go to Apple® App Store or the Android™ Market to download your MyChart Mobile App. Once downloaded, you will be prompted to search for URM C.

Check out <https://mychart.urmc.rochester.edu/mychart/> for more information.

From Peds to Adult Clinic

Donna Germuga, RT

We just had a very exciting revelation this week at the CF Center. For the first time, the number of adult patients with Cystic Fibrosis outnumbers the pediatric population! This is incredibly exciting because it means people living with CF are living longer, healthier lives! We talk a lot about transition to the adult center and why some people are very nervous and some extremely excited to be moving on and making more adult decisions for themselves.

One of the things you can count on as remaining unchanged in your transition is ME! I function as the RT on the pediatric and the adult side of the CF center. (Melissa the dietitian also goes back and forth between both centers) My role on the adult side is different in some ways from what is seen in pediatrics. I still do PFT's and talk a lot about airway clearance and nebulized medications, but also I function as the Adult Clinic Coordinator. I work with patients on usual items like prior authorization for routine medications and follow up from hospitalization or home IV tune up as well as more unusual items like how a patient traveling out of the country to a remote area will get all the medication and therapy they need to stay healthy while they are away.

One incredible opportunity I have been given is the chance to visit the lung transplant center at Columbia Presbyterian Hospital in Manhattan. This month, Tiffany (the adult center social worker) and I will travel to NYC to meet the team, observe pre and post transplant clinics and attend the team meeting where all potential recipients are discussed. We will try to contact some respite houses and other services while we are there to check out available resources. With so many people contemplating what transplant means and how the system works, this feels like a wonderful way to check it out, and at the same time put a personal face on the Rochester team so we have the best working relationship possible.



Saturday, June 8th, 5pm

Paradise Speedway in Seneca Falls

"Wings for CF" race will be supporting the Cystic Fibrosis Foundation

For more info check out our Facebook page:

https://www.facebook.com/Morganracing6?ref=stream&hc_location=stream

Your local Cystic Fibrosis Family Connection is in full swing and supporting YOU!

We had another successful basketball fundraiser raising close to \$12,000. Woohoo! This will fund our newly updated Hospital Help Packets, your pediatric clinic parking passes, community and educational events! Thank you to so many of you who helped with the event, raised funds, and helped us spread the word! See our website www.cffamilyconnection.org for more details, pictures, and even a video of our very own Brianna Collichio starting off the games with the national anthem. What a day!!

You can also visit our website to learn more about opportunities and activities in our community and beyond. Even better, while you're there, sign up on our email list (no spam, we promise) so that you'll be notified about important events and educational opportunities. Our Vests for Bosnia program, scholarship links, assistance, and educational opportunities are just a few of the things you will find.

CFFC, supporting our Rochester/Finger Lakes Community.

That means YOU :).

Hope to see or hear from you soon!



Cystic Fibrosis Family Connection

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Save the Date:
August 17, 2013, 7am, Geneseo, NY



For more information visit the CFF Rochester Chapter webpage at <http://www.cff.org/Chapters/rochester/>