

Inspire

Fall/Winter 2013

A newsletter for the Rochester Cystic Fibrosis Community

New Infection Control Guidelines

Clement Ren, MD

In our last newsletter, I outlined the Cystic Fibrosis Foundation's proposed infection control guidelines. We have been working hard since then to determine the best way to implement these guidelines. We have held multiple meetings amongst ourselves, AC-I clinic staff, inpatient staff and hospital administrators. We have carefully listened to and considered the opinions of our patients and families. We present our new protocols to you in this newsletter. We welcome feedback from all affected by these changes.

Patients and their families may be wondering why these changes are being recommended by the Cystic Fibrosis Foundation. We have long known that people with CF have chronic infection in their lungs because of the abnormal mucus and salt content in the lining of their lung tubes, but the specific organism in the lungs can vary. We also know that certain infections, most notably *Pseudomonas*, can be more serious. The changes in the CF Foundation recommendations come from new information over the last few years about how infections can be transmitted between CF patients. We have learned that bacteria can remain alive on medical equipment, such as lung function testing machines or blood pressure cuffs, for much longer than we previously thought. Bacteria can also remain suspended in the air for many hours. Based on this new information, the CF Foundation is recommending things like more intensive cleaning of medical equipment, minimizing contact between CF patients, and reducing exposure to potentially contaminated air.

We will never be able to completely eliminate the risk of new infections in CF patients. The bacteria that can infect CF patients are also in our environment, such as supermarkets and bathrooms. However, with these new changes we hope to minimize as much as possible the possibility that CF patients can acquire new infections in the clinic or in the hospital.

If you have any questions or comments about the new guidelines, please email us at **URCFPULMF@urmc.rochester.edu** and your question/comment will be routed to the appropriate clinic. Do not use this email address for urgent matters or to request appointments or medication refills.

Thank you for your cooperation.

Pediatric Outpatient Clinic Infection Control Protocol

Here is an update on the infection control changes we are making in the Pediatric CF Center (outpatient) in response to the new guidelines provided by the Cystic Fibrosis Foundation. We are asking for your participation to make these changes a success.

Recent studies suggest that if an individual with CF is coughing, the airborne droplets may travel up to 6 feet, further than was previously reported. With the new information, we want to take more precautions to minimize any potential infections between patients in our clinic.

- I. We ask that all individuals with CF put on a mask and do hand hygiene (hand sanitizer) when entering the AC-I outpatient clinic (any patient or family member with respiratory symptoms will be asked to do the same).
 - Masks and hand sanitizer will be available when you enter the clinic registration / waiting area.
 - Please wear the mask until you are in the exam room. Once in the exam room, you may remove the mask.
 - If you leave the exam room to go to the bathroom, please wear a mask and perform hand hygiene.
 - If you do need to cough, please cough into your elbow and/or a tissue and discard tissue in a hands-free receptacle. Then, perform hand hygiene.
 - If the CF patient is unable to wear a mask due to lack of cooperation, age, or other reason, consider these options:
 - i. If your infant or child with CF is in a car seat or stroller, you may drape a blanket over the top for protection.
 - ii. You may wait in the corridor outside of the entrance to the clinic after check in. Please let check-in staff know your location. While in the corridor, keep at least a 6-foot distance from others in the area.
2. Height and weight will be obtained in the current locations, but the remainder of your vital signs (O2 sat, blood pressure, etc.) will be obtained in the exam room.
3. Staff will wear gowns and gloves when we interact with you. We will perform hand hygiene and we will clean our stethoscopes before and after contact with you. Your exam room will be cleaned between patients.
4. Pulmonary Function Testing (PFTs) will be performed in the exam room.
5. Once your visit is over, we ask that you perform hand hygiene and wear a mask until you leave the building.



We will be implementing these changes in the Pediatric CF Center on AC-I starting in mid-December 2013. There may be a Friends of Strong Greeter in the waiting area offering you hand sanitizer and a mask. The CF Team is open to suggestions to make our process better. After a trial period, we will be introducing these changes to other outpatient areas within URM (like ENT, GI, Endocrine, radiology, etc.). However, we encourage you to regularly perform hand hygiene and wear a mask prior to that time when moving through other parts of the hospital.

Please do not hesitate to contact any of us on the CF Team with questions or concerns. We look forward to working with you to make these new guidelines a success. In the coming year, we also plan to create a brochure and a video walk-through of clinic to review these changes. Thank you for your cooperation!

Pediatric Inpatient Infection Control Protocol

Pediatric patients who need to be admitted to the inpatient unit for treatment are now usually admitted to 4-1400. The staff on this unit have been working very hard to help make your experience valuable for healing and for protection from infection. Because of the new guidelines to protect patients from getting more worrisome bacteria, we have started some new policies.

1. We are asking you (or your child) to wear a mask and do hand hygiene (wash or use alcohol based hand sanitizer) whenever you are on the unit outside of your patient room. This applies whether you are going in or out of your room, or visiting on the unit. This even applies when you are going to other areas of the hospital including to get PFTs, x-rays, cafeteria, gift shop or elsewhere.
2. Your doctors, nurses and other caregivers will do hand hygiene and wear gowns and gloves when we come into your room.
3. We will clean our stethoscopes before and after examining you.
4. It is important to remember that bacteria are spread up to 6 feet from you when you cough so please stay more than 6 feet away from other patients.
5. Child life staff will help to find activities to do in your room, but it will be helpful if you can bring activities that can be done in your room. This includes bringing school work so you don't get behind!
6. Child life and nursing staff will try to find time for you to get out of your room for activities, but this may occur at "off times" so you are not around other patients as much. This means that it is especially important to find a schedule that will work for you. You need to be able to do all your treatments every day as well.
7. Surfaces that you touch will need to be wiped down. This will be accomplished best with the help of parents as well as unit staff.

These policies are new and may take some getting used to. Let us know if you have ideas or other feedback to make this successful.



Adult Outpatient Clinic Infection Control Protocol

In January 2014, our approach to infection prevention will be visibly changing at the Adult Cystic Fibrosis Program at Culver Medical Group. Many of you are aware that the Cystic Fibrosis Foundation (CFF) has recently updated their guidelines for infection prevention and control. The changes were motivated by mounting reports of person-to-person transmission of bacteria between CF patients—also known as cross-infection—in healthcare settings. Our Program’s response has been informed by: detailed review of the CFF guidelines; consideration of the logistical, social, and emotional impacts of proposed policy changes; multiple discussions among the adult CF medical team, between the adult and pediatric CF medical teams, with other CF Care Center medical teams, and with members of our Adult CF patient and family community. While we have no reason to believe that our long-standing approach to infection control has resulted in even one instance of cross-infection between patients, we have decided to adopt substantial additions to our already serious protocols to prevent cross-infection.

First, we will continue our already stringent infection control program, which includes:

- Thorough disinfection of all surfaces of exam rooms between patient visits.
- Clinician hand and instrument (stethoscope) disinfection with bactericidal gel prior to and after exams.
- Scheduling patients with “complicated” infections at certain times and locations within the clinic.
- Pulmonary Function Testing (PFT) with a portable spirometer, disinfected between all uses.

The three goals of our enhanced infection control program, and the protections intended to meet them follow:

1. Limit time in the Waiting Area:

- We will move you from the Waiting Area to a clean and ready exam room immediately whenever possible, and otherwise as quickly as practical after check-in.

2. Limit potential exposures in facility common areas:

- A mask will be provided to all people with CF at check-in to protect you from exposure to germs from both CF patients and non-CF patients, AND to limit the spread of your germs to others.
- We ask that you wear the mask for mutual protection in all areas of Culver Medical Group, from entry to exit, except when you are in your exam room.
- Please clean your hands with bactericidal gel upon entering your exam room.
- Each time you leave the exam room (to check out or for the restroom), please clean your hands with bactericidal gel, and if you return to it, upon re-entering your exam room.

3. Limit potential cross-infection throughout the office visit:

- Program staff will wear a fresh, disposable gown each time they enter your exam room.
- Program staff will don fresh latex free exam gloves prior to examining you and again after examining you.
- Our famously friendly, affectionate Program staff will continue to express our affection verbally, but will no longer shake hands with or hug CF patients and their guests.

These important changes are intended to protect health and safety, and yet we recognize they will come with a cost. For example, we know from the experience of other CF Programs and from patient and family feedback that these efforts will for some reinforce a sense that they are “contaminated” and undesirable. We regret any hurt feelings that arise from our attempts to provide the most rational protection for all of our patients’ health and safety. We want everyone to whom we provide care—patients and loved ones—to feel safe and cared for in our hands. The experience at other CF Programs suggests ready acceptance of these changes. We are hopeful for the same, and we welcome your honest feedback as we all adjust to these changes.

Research Update

Mary Platt, RN & Nancy Nix, RN

Happy Holidays from your CF Center and Research Staff! As you know, there have been many exciting developments recently in the field of CF research. The approval of Kalydeco, almost two years ago, made by **Vertex** pharmaceuticals, has lead the way for more studies to continue to explore the safety and effectiveness of CFTR (CF gene) correctors and potentiators. These are the first medications which actually help correct the basic defect in cystic fibrosis and provide more chloride transport into cells where it needs to go. But, it is only for those with a specific gene (G551D).



Here at our CF Center and at Centers around the world, research in these and other areas continues to help advance CF care. While it is wonderful to have so many advances in the field of CFTR related medications, it is important that research in other areas continues to bring the best treatments to the front lines where they can be used to help those with CF be healthier. All of the clinical trials on CFTR and other drugs are done while research volunteers are still doing all of their routine CF related care. This reiterates the importance of those with CF to continue to do all of their treatments to gain and maintain the best lung function possible! Great Strides and other CFF fund raisers are vital to the CF Foundation's commitment to continued research.

The Genentech sponsored study, **IMPART**, that our Center participated in, to test the safety and effectiveness of the eRapid as compared to the Pari nebulizer for Pulmozyme, use ended this past summer. With the data from the study of 80 volunteer subjects from around the US, the sponsor and scientists were able to prove that this fast nebulizer is as effective and safe as the Pari nebulizer to use with Pulmozyme. It reduces the time to nebulize it to about 2-4 minutes, so it is a great time saver, and may increase adherence. Unfortunately, it is very costly, and most insurances do not cover it at this point. You may want to double check with your insurer to see if they cover it. Thank you to all of our wonderful pediatric volunteers and their families who made this study possible!

The **CFFC study** (Cystic Fibrosis Fibrosing Colonopathy study) has recently begun. This is a long term observational study sponsored by the drug companies that manufacture enzymes for CF. This study came about due to the rare occurrence of fibrosing colonopathy (FC-formerly known as colonic stricture aka narrowing of the large intestine, or colon) which has been known to occur in conjunction with enzyme doses that are very high. Since this is serious and usually requires surgery, it is important for them to assess the factors related to FC. There are no study visits, but observation via port CF by researchers after informed consent is given by study subjects (volunteers) or their parent. Some of the things that will clue us in on finding people for the study will be those who have had x rays revealing the possibility of FC after usual means to relieve abdominal pain have not worked.

Other CF studies besides the Vertex ones continue at our Center. We enter data after clinic visits into the **CF Registry** for those who have consented for us to do so. This data provides valuable information to see how our patients are doing over time, allows us to see how our Center is doing in relation to others, facilitates the development of quality improvement projects, and allows us to run reports and check patients' eligibility for research studies.

Our **EPIC observation** study of pseudomonas in CF continues and is in its tenth year. This has been a rich source of information for researchers who are trying to learn more about pseudomonas bacteria, why some people have pseudomonas bacteria, some don't, and the environmental and genetic factors that influence this. The annual surveys are currently being completed by 17 of our dedicated families and participants. We appreciate your time and efforts with this.

Visit CFF.org and view the research pipeline to get the most up to date information about CF research being done around the world at CF Centers like ours. Clinicaltrials.gov also has current research about CF and all other clinical trials. As always, we are grateful to our past, present, and future participants and their families. Call us if you ever have any questions. Mary Platt, RN 276-4123 Nancy Nix, RN 275-8580.

Along with the exciting developments in the treatment of CF that target the basic defect (Kaleydeco), there have been some recent medication changes you might be interested in for your pulmonary therapies. TOBI is now available as a generic. Your pharmacy may substitute generic if available to them. There is also another tobramycin inhaled formula called **Bethkis** that is formulated just a little differently and comes as a 4ml dose instead of 5ml resulting in a time savings over traditional TOBI.

The really exciting development for our patients 6 and over is the new **Tobi Podhaler**. It is a device that delivers Tobi as a dry powder in a capsule that can be done in less than 5 minutes and doesn't have to be refrigerated. We have available in clinic demonstrator models of the Podhaler so our patients who might be interested can experience it before we change over to make sure it is well tolerated.



We still have available the inhaled antibiotic **Cayston** (Aztreonam) for inhalation as you would use Tobi. It is done 3 times a day but the nebulizer is a special device that is small, portable and quiet, it also allows the treatment to be done in about 3-5 minutes (although some cleaning time needs to be allowed). You should discuss these treatments with your provider if inhaled antibiotics are on your treatment regimen and see if they are appropriate for you.

There has been some recent press on a vest called Afflo vest which is a self contained unit that the patient wears without hooking to a compressor. We trialed one here at the center and didn't have great feedback. The unit weighs about 10 pounds and doesn't seem to be as powerful as traditional vest units we have been using. There is also the issue of upgrade if the patient gains or loses significant weight. It would be up the distributor to decide how the upgrades are handled (and what the fee would be). The information we have also suggests that the lithium battery that powers the unit is only covered for one year after that the patient must pay for a new one.



As always, if you have questions or concerns about your respiratory therapies and airway clearance devices let us know and we will be happy to go over it to see if the vest is fitting properly and your other equipment is in good working order!

The board of CFFC would like to wish our community happy holidays.

We have so many reasons to both hope and be grateful looking ahead to 2014.

Check us out at www.cffamilyconnection.org to learn more.

Thank you for supporting us so that we can better support each other.



Patient Insurance Coverage of CF Medications

Michele Sanford and Christian Volk, Pharm.D. Candidates, and Angela Nagel Pharm.D., BCPS

In the ever-changing world of healthcare, it is important for patients to understand their insurance coverage, to ensure they get the care they need. Certain medications used to treat cystic fibrosis are considered “specialty” medications. These drugs may require the patient to use a designated specialty pharmacy. With MVP, BlueCross / BlueShield, and New York State Medicaid, the following medications require the use of a specialty pharmacy: Kalydeco, Pulmozyme, and Tobi. Medications like Cayston and Xolair depend on which insurance plan the patient is enrolled under. Many antibiotics, such as Azithromycin, often times can be picked up at a local (regular) pharmacy. It is important to bring a current insurance card to both the pharmacy and the doctor’s office, as insurance companies may update coverage in either January, July, or other times throughout the year.

Questions regarding specific coverage, like determining which specialty pharmacy you need to use, should be directed to your health insurance company. These numbers should be located on the back of your insurance card, but here are some of the most common companies in our area:

- ♦ MVP 1-800-852-7826
- ♦ BC/BS 585-325-3630
- ♦ Medicaid 1-800-541-2831

Please remember that we at the pediatric and adult clinics see patients from all areas of Upstate NY, from Buffalo to Albany and Watertown to Elmira, so we can’t possibly keep up on all the different insurances our patients have. Knowing your insurance will help us to get the medications that you or your loved one needs faster.

Vitamin Update, Again

You now have FOUR CF vitamins to choose from! AquADEK has been around the longest and is most likely covered under your insurance. Call your insurance to see if the new ones are covered:

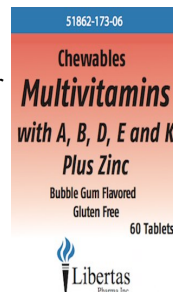
ChoiceFul Vitamins

- Chewable, Softgel and Liquid Drops
- www.choicefulvitamins.com
- Offered Free at Foundation Care Pharmacy when you fill other CF meds there.
- Order online from Foundation Care or IVSolutions. \$15 for a bottle of 60



Libertas Pharmacy ABDEK Plus Zinc

- Chewable, Softgel and Liquid Drops
- www.abdeks.com
- Offered Free via Creon CFCareForward
- Order online or have your pharmacist order it for you



MVW Nutritionals Complete Formulation

- Chewable and Softgel
- mvwnutritionals.com
- Offered Free via Creon CFCareForward
- Order online or have your pharmacist order it for you. \$29 online for bottle of 60



The Affordable Care Act (ACA) and You

Marcy Odell, LMSW

For many people with cystic fibrosis, the ACA offers some new health insurance options and protections.

Several of these changes may be helpful for the CF community:*

- ◆ Insurers will no longer be able to deny coverage based on a pre-existing condition like cystic fibrosis.
- ◆ Since 2010, young people are able to stay on their parent's health plan until they turn 26.
- ◆ Beginning in 2014:
 - ◇ New state marketplaces will help consumers compare and purchase different health plans.
 - ◇ Lifetime and annual limits will be eliminated for essential health benefits on many plans.
 - ◇ Several states will expand Medicaid to include more low-income adults.
- ◆ Beginning in 2015:
 - ◇ Out-of-pocket expenses will be capped.

For more information go to: <http://www.cff.org/LivingWithCF/Insurance/ACA/>

Or to access the healthcare marketplace go to: <https://www.healthcare.gov/>

Or contact Marcy Odell, LMSW, at marcy_odell@urmc.rochester.edu

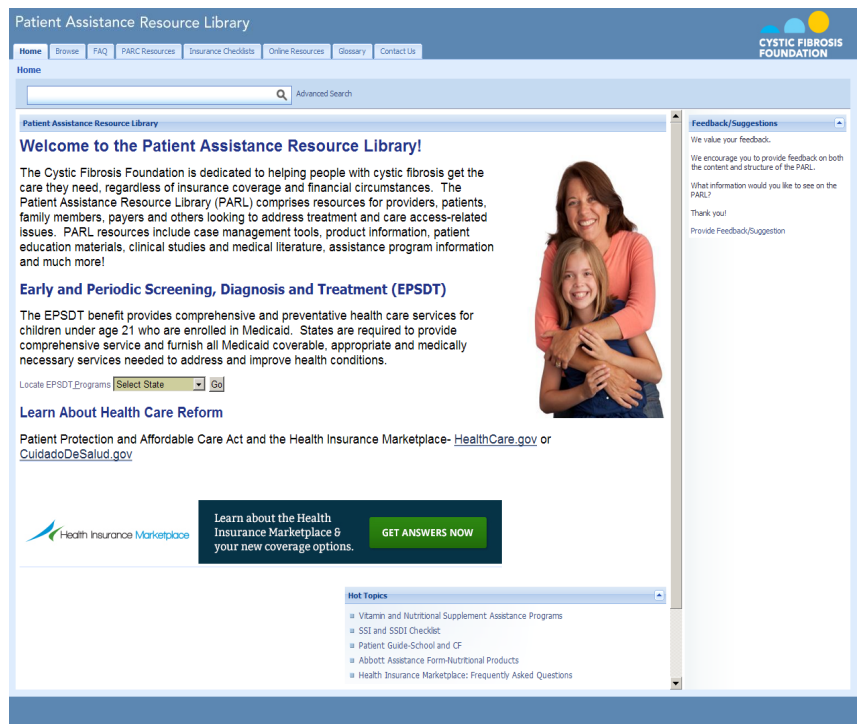
Patient Assistance Resource Library

To help you navigate the complex world of health insurance and healthcare coverage, the Patient Assistance Resource Library offers a range of useful information:

- ◆ Sample letters of medical necessity and prior authorization;
- ◆ How-to guides to navigating common insurance obstacles;
- ◆ Templates for insurance appeals, exceptions and reconsiderations;
- ◆ CF care guidelines;
- ◆ Evidence-based medical journal articles

For more information go to:

<http://www.cff.org/LivingWithCF/AssistanceResources/Library/>



The screenshot shows the Patient Assistance Resource Library website. The header includes the title 'Patient Assistance Resource Library' and the Cystic Fibrosis Foundation logo. A navigation menu contains links: Home, Browse, FAQ, PARC Resources, Insurance Checklists, Online Resources, Glossary, and Contact Us. Below the menu is a search bar with an 'Advanced Search' link. The main content area starts with a 'Welcome to the Patient Assistance Resource Library!' message, followed by a paragraph explaining the library's mission. It then lists resources for 'Early and Periodic Screening, Diagnosis and Treatment (EPSDT)' and a section to 'Learn About Health Care Reform' with links to 'Patient Protection and Affordable Care Act and the Health Insurance Marketplace- HealthCare.gov or CuidadoDeSalud.gov'. A sidebar on the right contains a 'Feedback/Suggestions' form. At the bottom, there's a 'Hot Topics' section with links to 'Learn about the Health Insurance Marketplace & your new coverage options.' and a 'GET ANSWERS NOW' button.

When You Can't Get Enough Calories

Melissa Berry, RD

We all know the recommended diet for those with Cystic Fibrosis is high fat, high calorie. And we all know that people with CF have a higher calorie demand than those without. Some of you have experienced that it is often difficult to meet such high calorie needs in good health, let alone meeting them when suffering an exacerbation.

The first step when looking into difficulty gaining weight is to make sure you are taking your enzymes appropriately. The next step is for your provider to make sure you are on the correct enzyme dose, which is based on your weight. Sometimes trying a new enzyme helps. You may also need to be evaluated for reflux or CF Related Diabetes, both of which can hinder weight gain efforts. Your dietitian will work more closely with you to add in higher calorie foods and ask you to try supplements such as Pediasure, Ensure, Boost, and Scandishakes; or try adding a powdered calorie supplement to your foods. Sometimes calorie counts help you to see how many calories you are actually eating so you can get a better idea how much more you or your child need. But sometimes, despite doing all of the above perfectly, it is just too difficult to meet the body's demands for energy to keep your child growing properly or you from losing weight. This is when we start thinking about placing a gastrostomy tube, or G-tube.

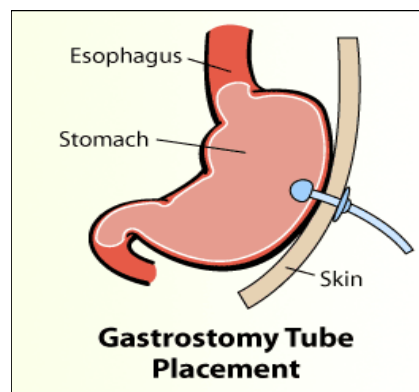
The diagram below shows that a g-tube is placed directly through the abdominal wall into the stomach, and held in place by a balloon on the inside and a round disc on the outside. For the first few weeks, a longer tube is attached while the site heals. After this, a smaller and less noticeable "button" can be placed (below, left).

Tube feeding is usually done overnight and patients can get up to 2000 additional calories while sleeping! Sometimes, if a person's appetite is low or they feel unable to eat due to coughing or illness, feedings can be done during the day. Some children have a school nurse give them a feeding during the school day.

Thinking about a g-tube is difficult. Many think that needing a g-tube is a sign of advancing disease and that they may not get better. This is not the case! Children usually need it because they simply cannot eat as many calories as their body requires to help them grow normally and sustain good health. Some people who experience a severe exacerbation or repeated exacerbations and have lost a lot of weight need the extra calories overnight to help them regain weight and regain their health. Some will get a g-tube to help with weight gain before a lung transplant. Women with CF can get overnight feedings to help them gain weight during pregnancy. G-tubes are typically used for several months to several years, so they need not be permanent. But whatever the reason, it is a scary step to take and requires a lot of thought and planning and commitment. We have patients of all ages with g-tubes that would be happy to talk with you if you are considering a tube for yourself or your child.

You might benefit from a G-tube if:

1. You are unable to take in enough calories to gain or maintain weight
2. You have lost your appetite or have noticed it is not as good as it used to be.
3. You often vomit from coughing and/or have severe heartburn
4. You are unable to regain weight that was lost when you were sick
5. You are trying to reach a good weight for lung transplant






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Two decorative circles, one solid light blue and one outlined light blue, are positioned to the left of the "Save the date!" box.

Save the date!

March, 2014 Annual Kit Taylor Lecture featuring
guest speaker Dr. John J. LiPuma, Director of the
Division of Pediatric Infectious Diseases at the
University of Michigan

May 17, 2014 Great Strides Geneva

May 18, 2014 Great Strides Rochester

June 1, 2014 Great Strides Hornell

For more info and to register for Great Strides go to Fightcf.off.org