University of Rochester Medical Center
Cystic Fibrosis Center

Inspire

Spring/Summer 2011

Treatments...ugh

Dr. Rob Horowitz

Why does my CF provider team push me to spend so much time and energy doing daily treatments that don't make me feel any better, and sometimes make me feel worse?

Important question! I confess, sometimes I am so eager to optimize my patients' daily treatments that I don't adequately discuss their important day-to-day impact, both good and bad. But this discussion is vital, because daily treatment plans for most people with Cystic Fibrosis are complicated and time-consuming, and I can't expect adherence to such a regimen without fully discussing both its expected benefits AND its challenges and side effects

For the vast majority of our patients with CF we recommend a variety of nebulized medications and airway clearance techniques (ACTs), including exercise, all of which require a significant expenditure of time and effort. And our questioner is right: These daily treatments often do NOT make people feel immediately better, and, in fact, they make some people feel temporarily worse. Why do we push our patients to perform such demanding, unrewarding, and sometimes unpleasant treatments?

In essence, daily CF lung treatments (please note: we are here discussing routine, daily treatments, not acute treatments during exacerbations, like "tune-ups", for example) are intended primarily to help you keep your lungs as healthy as possible for as long as possible. Many people do enjoy immediate benefit from these treatments, like the clearing of stubborn mucus and breathing easier. When this happens, the reward of feeling better tends to reinforce adherence to these treatments. Unfortunately, many people, like the questioner, do not enjoy such immediate rewards, but rather experience only the time imposition and sometimes unpleasant treatment effects. In this case, our recommendations may appear irrational, if not downright insensitive! However, our motivation is to help you enjoy long-term treatment benefits, even if the process is cumbersome and sometimes uncomfortable.

Let's consider Hypertonic Saline as an example. Some people are thrilled how this medication enhances their cough and airway clearance. Indeed, small studies suggest this may provide fairly rapid, modest improvement in lung function. However, many people feel no immediate effect, while others are irritated by side effects (hoarseness, voice changes, chest tightness, wheezing, increased cough, and sore throat). And most are acutely aware of the time demands: Preparing, nebulizing, and cleaning up after this treatment twice daily takes about twenty to thirty minutes total for most people, a significant time drain—over the course of a year this amounts to over 100 hours spent on just one medication! How do we justify this recommendation? Hypertonic saline offers two main benefits: it does, in fact, help many people immediately clear some mucus from the lungs; in addition, and probably more importantly, we believe this medication improves the lungs' fluid lining to a normal depth, essentially reversing CF's hallmark thin, dehydrated airway surface liquid depth, such that the lungs can clear the mucus and pus that otherwise accumulates in CF lungs. This effect is not likely to make one feel immediately better, and indeed, as stated above, it makes many people feel temporarily more uncomfortable. But over months to years, we have reason to believe this can limit cumulative lung damage, and thereby protect lung function and overall health.

Many of the other daily treatments your provider team prescribes are similar: They impose an increased burden of time and effort, sometimes with little detectable immediate improvement and often moderate short-term discomfort, for a long-term payoff of better preserved lung function for a longer time period. We are eager to help you understand both the immediate and the long-term, deferred benefits of the treatment regimens we recommend. And we are committed to collaborating with you in determining which of the growing number of available treatments are most likely to provide you both immediate and long-term benefit, recognizing always the impact of the treatment on your daily life.

We Need You!

Do you have ideas for making your clinic visits easier? Your hospitalizations more manageable? All are welcome to participate in the CF Family Advisory Board (FAB). Our aim is to gather your thoughts and ideas to help make our center better for you! The next FAB Meeting will be *Wednesday, June 22, 2011, at 6pm*. Location to be determined. Please contact Marcy Odell at 275-9105 or email her at Marcy_Odell@urmc.rochester.edu for more information.

Inside this issue:

Managing the Behaviors of Children with a Chronic Illness

edited by Marcy Odell, LMSW

"If we picture an individual from birth as a paddler in a river, we can see that his or her parents do all the paddling initially but gradually teach the child skills so that by late adolescence, the child is able to take over the canoe with some coaching from the shore. To continue this metaphor a little farther, there are a variety of obstacles in the river; some that you can see coming such as turns or rapids, but occasionally there is a submerged log that calls upon the skills of all the paddlers in the boat. The diagnosis of a chronic illness in a child can be like a submerged log. Some need help from each other to get back in the canoe. With a few paddling lessons they'll be more prepared next time. Perhaps the caregiver(s) will have to stay on board a little longer but our paddler will eventually become the captain (Brownlow, 1993, 1)."

A major task parents of a chronically ill child face is the responsibility of helping their child cope with his or her illness. Some suggestions are educate yourself about your child's illness, explain the illness to your child, help your child deal with his or her feelings about the illness, don't be afraid to discipline, and give your child responsibilities. Other suggestions include maintain family routines as much as possible, prepare your child for medical procedures, help your child lead as normal a life as possible, take care of yourself, prepare your child for the reactions of others, be mindful of what your child can overhear, let others help, give your child some choices, help your other children cope, and work closely with your child's school.

Parents should try as much as possible to treat their chronically ill child like any other child. At the same time, they need to take into consideration their child's illness and any special needs that he or she may have. This can be quite a balancing act for parents. *It is very important for parents to encourage participation in various activities that involve other children of the same age*.

A chronically ill child demands a lot of parental attention. It is no wonder that brothers and sisters often feel jealous, angry, and lonely. Siblings also worry about their ill brother or sister, worry about their parents, and worry that they might get the disorder, too. Therefore, it is important for parents to spend time with their other children to provide a sense of security and to help them cope. Parents should explain the illness to their other children and try to get them to ask questions and to express their concerns. Parents need to maintain open lines of communication with all of their children. It often helps children feel like a more important member of the family to help care for their sibling in some limited way. Whenever possible, parents should try to schedule and spend individual time with their other children to help them feel important and loved.

Regardless of the condition, children and adolescents have similar feelings about chronic illness-apprehensions, rage, self-pity, and bitterness. The young child, unable to understand why the sickness has occurred, may assume it is a punishment for being "bad." He or she may become angry with parents and doctors for not being able to cure the illness. The youngster may react strongly against pampering, teasing, or other attention. Uncomfortable treatments and restrictions in diet and activity may make the child bitter and withdrawn. It may take a little while to accept the situation, but it will happen.

Parenting a chronically ill child is a challenge. Many parents are reluctant to set limits with their chronically ill child. However, just like any other child, the chronically ill child needs discipline from his parents. Discipline provides children with structure and security, which is very reassuring to a child. Adequate discipline helps children learn to control their own behavior, too. Parents should make sure that discipline is consistent, both between parents, and from day to day with individual parents. Children need to know what to expect from their parents. Parents should also make sure that other family members and anyone else who cares for their children use consistent discipline, too. Recommended discipline techniques include praising appropriate behavior, using time-out with young children, and restricting privileges for older children. It is important to consider the development age of the child when examining effective ways in managing behaviors.

Infants (birth to 12 months)- they need schedule around feeding, sleeping, and play or interaction with others. The schedule helps regulate autonomic functions and provides a sense of predictability and safety. They should be allowed to develop some tolerance to frustration and the ability to self-smooth.

Early toddlers (1-2 years)- it is normal and necessary for toddlers to experiment with control of the physical world and with the capacity to exercise their own will versus that of others. Ways to manage the behavior of early toddlers is to remove the child or the object with a firm no, or another way is to provide a brief explanation ("no,hot") and redirecting the child to an alternative activity. Early toddlers are very susceptible to fear of abandonment and should not be kept in time-out away from parent.

Late toddlers (2-3 years)- the caregiver should continue to supervise, set limits and routines, and have realistic expectations of the child's achievement capabilities. An example is when a toddler is having a temper tantrum in a public place. Remove the child from the place of misbehavior. Hold the child gently until the toddler gains control. Give a short verbal instruction or reassurance followed by supervision and an example.

Preschoolers and kindergarten-age (3-5 years)- most children are able to accept reality, limitations, and act in ways to obtain others' approval and be self-reliant for their immediate needs. They have not internalized many rules, and their judgment is not always sound. Time-out can be used if there is loss of control on the child's part. Redirection or small consequences related to and immediately following the misbehavior are other alternatives.

School-age children (6-12 years)- conflicts may arise at this stage because of the child's increasing independence. Children at this stage tend to act autonomously, choose their own activities and friends and, to some extent, recognize other than parental authority. Parents should continue to make the important decisions, because school-age children cannot always put reasoning and judgment into practice. Acceptable means of managing behavior are withdrawal or delay of privileges, consequences, and time-out.

Managing Behaviors *con't*

Adolescents (13-18 years)- conflicts frequently ensue because of the adolescent's adherence to the peer group, challenging family values and rules, and distancing from the parents. The parent can meet these challenges by remaining available, setting rules in a non-critical way, avoiding belittling the adolescent, and avoiding lectures. Contracting with the adolescent is a useful tool. Many adolescents do want parental guidance and approval. Parents should ensure that the basic rules are followed and that logical consequences are set and kept in a non-confrontational way.

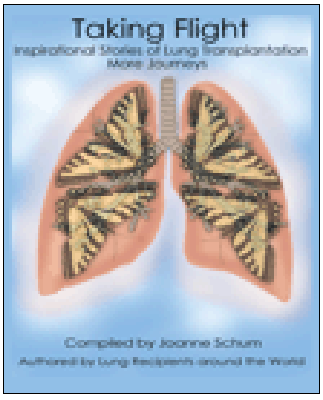
Some ways to manage your child's behavior:

- Tell your children you are pleased when they behave well and cooperate
- Avoid only pointing out mistakes
- Children learn how to behave by watching the adults who care for them
- Avoid power struggles by using distraction or by giving choices
- Spend time every day giving each child your complete attention
- Do something fun: read a book together, take a walk, play catch, or do some other simple activity
- When a behavior can't be ignored, use disciplines appropriate for your child's age. For toddlers (18 months to 3 years), distraction or redirection works best. For preschoolers (3-5 years), use time-outs-one minute for each year of age.
- Talk about angry feelings with your children. Try to sort out the reasons for these feelings in yourself, and in your child.
- Say "**I LOVE YOU**" to your children. Say it a lot.
- ***Remember: Parenting is hard work, but it's worth it!***

This article was obtained from the Center for Effective Parenting http://www.parenting-ed.org

A Star Among Us: Joanne Schum

Our very own Joanne Schum proudly announces the publication of her newest book:



"Taking Flight: Inspirational Stories of Lung Transplantation-More Journeys"

What do Medical Professionals say about "Taking Flight – More Journeys":

"It is an honor to be asked to write an Introduction to the second edition of Joanne's anthology of inspirational stories about patients who have received lung transplants, and encouraging others who embark on this journey."

~Dr. Thomas M. Egan, Professor of Surgery, University of North Carolina at Chapel Hill, Chapel Hill, North Carolina

"I concluded that it was a good thing that lung transplantation might transform the lives of a few but would give hope to all – that there was a potential treatment. One recipient told me, 'I have no more rainy days. Every day is sunshine.'"

~Dr. Joel Cooper, Thoracic Surgery, Chief of the Division of Thoracic Surgery at the University of Pennsylvania Health System, Philadelphia, Pennsylvania - Regarded as, "The Father of Lung Transplantation"

"I think Einstein would have enjoyed Joanne's books."

~Dr. Marshall Hertz, Director, Center for Lung and Health, Medical Director, Lung Transplantation Program, University of Minnesota Medical Center, Minneapolis, Minnesota

What does the public say about, "Taking Flight – More Journeys":

"My sister bought 'Taking Flight' for my Dad. Last night we sat through our second 'dry run' and Joanne's book was like the bible. My Mother and Father do not go anywhere without it."

~Addie Benton Poudrier

What does Joanne say about, "Taking Flight – More Journeys":

"Like the flutter of butterflies – a fresh gathering of lung recipients share the success of their 'Gift of Life' from an organ donor. Along with, stories from organ donor families, women who gave birth after lung transplantation, and the medical professionals whose expertise allow a future for us all."

~Joanne Schum

To Purchase:

Mail a $14.00 Money Order made out to Joanne Schum to the following address:

Joanne Schum
1104 Bay Road
Webster, NY 14580

Joanne would be happy to autograph the book for you, just include a note with your money order indicating you would like it autographed.

Cystic Fibrosis Cycle for Life

by Chris Kvam

I've been asked to write a follow-up article to my piece about exercise that appeared in last fall's newsletter. This is exciting, and particularly challenging, as I've never been asked for a follow-up on the same topic before! While I've had a full winter and spring of cross country skiing and cycling, I'd like to talk more about putting some of my advice in last fall's article to work.

I ended my last article encouraging others with CF to start incorporating exercise into their daily routines. I also wrote about how scheduling events on the calendar helps motivate me to keep a high level of fitness year round. So, here's a chance to pick something to train for, a date to circle on the calendar, and something to get your friends and family involved in and excited about! On August 20th, the Rochester CFF will host its first annual Cystic Fibrosis Cycle for Life, a one day bike ride through the western Fingerlakes region (see the Rochester Chapter page for details and registration). The ride will start and finish at SUNY Geneseo, and offer 65 and 35 mile routes. The ride will be fully supported with rest stops every 10 miles or so, support vehicles if anyone needs to call it a day early, mechanical support, and a great picnic lunch hosted by SUNY Geneseo. The route will be somewhat challenging (moderately hilly), but rewarding – with great views, downhills, and a sense of accomplishment.

The ride website has suggestions for training for a 65 mile ride over 2-3 months, which gives you plenty of time, assuming this is printed sometime in June! If anyone needs any guidance regarding training, equipment, etc. feel free to contact me at the email shared below.

I've been spending quite a bit of time organizing this ride for a few reasons. First of all, I love cycling, and I have the desire to share how important cycling and exercise in general have been to my efforts to live life fully with Cystic Fibrosis. Secondly, I am acutely aware that my life and abilities would have been very different if I didn't have access to the drugs developed in partnership with the CF Foundation over the last 20 years. It is my hope that this ride both celebrates how far we have come in my lifetime, while recognizing how much work remains to be done, and fundraising towards that ongoing effort. Lastly, although I'm relatively new to the area, I think that Rochester needs some CF events right now. The changing chapter organization is difficult, but the loss of office staff does not change the potential for people in the Rochester CF community to put on events, raise money, and support each other.

So, here's your chance! Ride for yourself, for a loved one, for friends, and for a great cause. If you're not interested in riding, we will need volunteers to set up and staff rest stops, cheer riders at the finish, and organize a silent auction for during lunch after the ride. If you have any questions and/or are willing to volunteer your time and efforts, feel free to email me, Chris Kvam at cmkvam@gmail.com.

Cystic Fibrosis Family Connection Gets a Boost

by Kori Tolbert

Many of you who have been involved in the Rochester CF Community are also familiar with the Cystic Fibrosis Family Connection, CFFC. For those of you who aren't familiar with CFFC yet, let me introduce you. This important organization has supported children, adults, and their families dealing with a life with CF since 1993. Many of the original participants were parents of children with CF and had worked closely with the Rochester Chapter of the CF Foundation prior to forming CFFC. They saw the CF Foundation's (CFF) focus seemed to be moving exclusively to support the medical aspects of dealing with CF- research, the pharmacy, etc. The great advances in medical treatment that the CF Foundation has sponsored are incredibly important in our battle to prolong lives and eventually cure CF. However, the intention with the creation of CFFC was to fill the gap in the support that CF families need with the ongoing, day to day care of patients. It was clear to them that CF patients and their families needed help dealing with the implications beyond direct medical care. So, the Cystic Fibrosis Family Connection (CFFC) was formed.

The intention of the CFFC was to provide education, financial assistance, and general support for our local families. CFFC has helped with things like hospital assistance packets to help pay for phone, TV, and computer use for in patient stays, parking passes, a toy closet, CF Family Days and community education, emergency financial assistance, and other support projects that have assisted us in living and dealing with the complications of a "CF life". Recently the organization has been struggling for financial and community involvement and had come to a place where they were about to close the doors. When my boyfriend, Rick Doherty, and I learned that CFFC was coming to a place where it would no longer exist, we called to see what we could do. At one point Rick had suggested the possibility of a basketball tournament to raise funds and awareness. Unfortunately I was dealing with a CF flare up at the time so it wasn't something I could do right away, but we knew we wanted to help when we could.

A few months later one of my dear friends, Peter Lunn, who also had CF, passed away due to complications after his lung transplant. Peter is originally from Jordan, outside of Syracuse, but he went to our Rochester clinic as a young teen and into adulthood. That's where we met actually, as kids on 1400 at Strong. After his passing I knew I wanted to do something to honor my beautiful friend. I wanted to do something special, something that said, YES TO LIFE, in my friend's honor. And then it came to me, PETER LOVED BASKETBALL! I remember talking with him after his lung transplant and one of the things he was most excited about was the possibility of playing basketball again. (Which he did, along with a trip to Vegas, and more.) Peter benefited from some of CFFC's programs directly. I'm sure he would be happy to know that he was a part of reviving the organization; making sure other children, and adults, with CF (and their families) could benefit from CFFC for years to come. It felt like a good fit. And so, the plans for the CFFC 3 on 3 Basketball Tournament in honor of Peter J Lunn had begun.

CFFC *con't*

The Tournament took on a life of it's own and on March 26th from 1:00 in the afternoon until about 5:00 we had over 40 teams of all ages competing for their chance to win their division's trophy. We had lots of delicious baked goods, snacks, pulled pork, pizzas, and drinks donated for the event. In the end, we were able to give CFFC the significant boost that it needed earning over $7,500. This tournament was a big chunk to take on, but CFFC has been there to help when our CF Center and families needed support. It feels good to help ensure it's existence. In a way, I think it not only helped CFFC, but also me to move forward. I found Peter's loss incredibly hard; the tournament became a way to connect me to this world and Peter. It was a beautiful event and Rick and I are so very grateful for those that donated their time, money, and energy to make it a success.

Our group of volunteers at CFFC are currently working together to make sure to provide help that will reflect the feelings and needs of the current Rochester CF community. We are looking for both community support and feedback. If you have any ideas for community support or interest in joining our CFFC volunteers please contact me at ktwildflower@hotmail.com or 585 350 9121. Donations can still be made and mailed to: CFFC, PO Box 93328,Rochester NY 14692

Learning at Home: An Option for Families of Children with CF

Our children have never gone to daycare or public school. Yet, they are happy, healthy, intelligent, and socially well adjusted. As parents of children with Cystic Fibrosis, we clearly understand the obstacles to maintaining good health. We have found that homeschooling helps to create less stress in our daily lives, while allowing the opportunity to monitor and assess our children's health. We have homeschooled for years, and found that our children being "schooled" at home has provided us the wonderful opportunity to meet the demands of this disease.

Homeschooling provides us with adequate time to complete airway clearance and nebulized treatments each and every day. Additionally, it teaches our children the importance of habits and routines. It ensures that our children receive and consume healthy high caloric meals and snacks. It enables us as parents to determine the correct number of enzymes to be given without leaving it in the hands of someone else. It creates opportunities for our children to sleep in or rest when needed. It allows us the opportunity for flexible scheduling (which is great for CF appointments or participation in clinical trials). Homeschooling allows for less exposure to "illnesses of the day" that are common in daycare or public school settings.

Homeschooling is not for every child or every family. It is easy to get started, and easy to stop. Whether you consider homeschooling for weeks, months, or years, you can rest assured there are a significant number of resources available to homeschoolers and their families.

Please feel free to contact me via e-mail with any questions you might have. My e-mail address is bnsbch@yahoo.com.

Nutrition Notes

Melissa Berry, RD

New guidelines were released in December 2010 outlining the screening, diagnosis and treatment of Cystic Fibrosis Related Diabetes (CFRD). CFRD is considered the most common co-morbidity in CF and affects nutritional status as well as pulmonary status. CFRD is not the same kind of diabetes found in someone without CF. It is treated, diagnosed, and managed differently. There are 3 main causes of diabetes in CF patients:

1) Insulin Deficiency: Insulin is made in the pancreas. People with CF make less insulin due to scaring on the pancreas from the thick mucus that collects there.

2.) Insulin Resistance: This occurs as a result of chronic or acute underlying infections that often accompany CF. When this occurs the body does not use the insulin as effectively and needs more insulin to get the same job done.

3.) High Cortisol Levels: High cortisol levels interfere with the action of insulin and causes insulin resistance. Cortisol levels increase in our bodies when we are stressed and also by taking drugs called corticosteroids that many CF patients take to treat lung infections.

CFRD is diagnosed by a blood test called an Oral Glucose Tolerance Test (OGTT). The new recommendations are to begin annual testing at age 10. The test is ordered by your physician and is carried out in the lab. It is a two hour test that requires an 8 hour fast. Upon arrival at the lab, a fasting blood sugar is taken. Then the patient will drink an 8 oz super sugary solution called Glucola. The patient must sit and rest for the next two hours when the final blood draw takes place.

Your doctor will let you know the results of the test. If you have CFRD then you will be referred to an Endocrinologist for further diabetes management. Treatment includes daily monitoring of blood sugars as well as insulin injections. Unlike Type 1 and Type 2 diabetes, a special diet is not recommended. The high calorie, high fat, high protein, high carb diet is still the recommended diet for those with CFRD.

CFRD Highlights

- Average age of onset 18-24 years
- 20% of adolescents and up to 50% of adults have CFRD
- Can cause weight loss and decreased pulmonary function
- Recommend annual screening beginning at age 10.
- Treated with insulin

Nurses Nook

Bridget Platania, PNP and Pat Lamarche, PNP

We often answer questions about the most cost effective way to fill prescriptions. Every insurance policy is different and we advise you to check with your specific insurance company for the most up-to-date information.
In general, a course of oral antibiotics or a 30- day supply of a maintenance medication is filled by your local pharmacy. It is beneficial to maintain close communication with your local pharmacist as some CF medications may need to be special ordered. Some insurance companies offer a 90-day supply option which may be less expensive. A Customer Service representative (phone number often listed on the back of your insurance card) may be helpful in answering questions specific to your policy. In addition, here are some alternatives specific to CF medications:

- **The CF Services Pharmacy**
  - Run by the CF Foundation
  - http://www.cfservicespharmacy.com
  - Stocks specific CF medications which are almost always in stock
  - Shipped to your door
  - Patient/Family must sign up (either on-line or via phone at (800) 541-4959)
  - Typically, if this pharmacy fills your inhaled medications (TOBI, Cayston, or Pulmozyme) you can receive hypertonic saline for inhalation at no charge
- **Foundation Care Pharmacy**
  - http://www.foundcare.com/cysticfibrosis.aspx
  - Stocks specific CF medications which are almost always in stock
  - Shipped to your door
  - Patient / Family must sign up (either on-line or via phone at (877) 291-1122)
  - See website for special offers (i.e. if you receive enzyme supplements through this pharmacy you can choose to receive either vitamins or hypertonic saline free of charge)

The Pediatric CF Center does not recommend one pharmacy over another and again, encourages you to contact a customer service representative to discuss your specific plan.

Social Work Bites

Tiffany Passalaqua, LMSW

Summer time is a great time to gather up the family and friends and head out on a vacation. Make sure to fit your therapies into vacations and other activities in life. Your health should remain number one. No trip is fun if you're not feeling well. Here are some tips for making your vacations memorable, healthy and safe:

- Eat well, Stay hydrated, sleep and stay compliant. It's easy to skip meals and therapies when on vacation but if you can stay in your routine as much as possible, it will lessen the stress that traveling causes your body.
- Check with the doctor and make sure your prescriptions are full. Bring 3-7 days extra just in case! If you are flying- keep all your medications in your carry-on and keep them in their original prescription bottles. It would also be helpful to have a note from the doctor with your diagnosis and medication list.
- Find the nearest CF center to your destination and tell your companions what to do in case of an emergency.
- Plan "rest" days before and after your vacation to help your body recover. Whenever you get out of a routine- your body will respond and want REST! Listen to your body and slow down and rest. Don't be afraid to ask for a wheelchair or scooter at the airport or amusement park.
- Think about airway clearance-is your vest too heavy for the plane? Can you use your acapella for a week? Or rely on exercise or cupping? Talk to your doctor on what is best for you.
- Consider luggage on wheels or shipping things ahead of time to lighten the load you will need to carry with you.
- Make sure you have antibacterial wipes or gel and use them frequently!
- Bring a water bottle and extra snacks (high carb snacks if you are diabetic).
- Consider taking emergency medications with you like antibiotics and constipation relief meds. Ask your doctor what would be appropriate for you.
- Bring insurance information and identification with you and know what your policy covers for out of area charges.

Remember to be flexible and have reasonable expectations for what you want to accomplish. So you might not make it as far as you planned if you are driving. You might not get to see everyone or go everywhere on your list. What is important is having fun, relaxing, and most of all, staying healthy! And don't forget to bring pictures to your next clinic visit!

Research Corner

Mary Platt, RN and Nancy Jenks, RN

We first would like to let you know about our new Research Nurse, Nancy Jenks. Nancy started with our Center in November after our Research Coordinator, Marissa Dixon, left to pursue her masters' degree in public health. Nancy has experience with several pediatric studies as well as adult clinical trials. She came to us from the Department of Pediatric Infectious Diseases where she worked as a research nurse and is enjoying working with the Pediatric Pulmonary Group and CF patients.

iCARE

A lot of you already know about one of our studies currently in progress called iCARE (I Change Adherence and Raise Expectations). We have enrolled 18 of our patients. Asking people to do more treatments and take more medications during the course of their busy days is very challenging but very important. As Dr Alexandra Quittner, the lead Principal Investigator of the study at the University of Miami notes, "Patient non-adherence is a large contributor to poor health outcomes and increased clinic and hospital visits." The goal of iCARE as its name implies, is to help you and our center staff learn more about how to help subjects (patients in the study) find ways to incorporate all of their care (airway clearance, supplements, and medications) into their days. We are working to gain tools that will help us help empower our teens to make small changes toward leading to bigger ones.

This study also incorporates procedures that are helpful in caring for all of our patients, not only the ones who have consented to be in the study. For instance, one of the "skills checks" involves asking the participant in the study and his/her family to bring all of their respiratory equipment (vest, nebulizer, compressor, spacer) and demonstrate its use with Kim or Donna. Another part of the study involves problem solving sessions with "brainstorming" in which the teen in the study leads a discussion and decides what she/he would like to improve on. A goal is set by the teen and the specifics are discussed. There is also follow up by phone to discuss the goal chosen.

We look forward to continuing to work on the study. Thank you for all who have enrolled and to all of our patients, please give us feedback on how we can you help with adherence.

Vertex 809-102

Many of you probably have heard about the Vertex studies. Vertex is a compound that works on the CFTR channel (cystic fibrosis transmembrane redactor) Vertex 809-102 is a phase two study looking at the safety and effectiveness of the study drug, VX 809 when taken alone and in combination with the study drug VX 770 in people with CF who have two copies of the delta F508 CFTR gene. The following information is quoted from NIH.gov: "Disease-causing mutations in the CFTR gene alter the production, structure, or stability of the chloride channel. All of these changes prevent the channel from functioning properly, which impairs the transport of chloride ions and the movement of water into and out of cells. As a result, cells that line the passageways of the lungs, pancreas, and other organs produce mucus that is abnormally thick and sticky. The abnormal mucus obstructs the airways and glands, leading to the characteristic signs and symptoms of cystic fibrosis."

Below are some of the criteria for the study as found on www.clinicaltrials.gov and there is more info at www.cff.org search word "vertex." We expect to enroll people in early fall 2011. Please contact us at 275-8580 (Nancy) or 276-4123 (Mary) if you have an interest in the study. Since it is a phase two study, it is very involved and includes some all day visits and enrollment is limited.

Eligibility Criteria:

- 18 years and older
- Confirmed diagnosis of CF
- Must have 2 copies of F508del-CFTR mutation
- FEV1≥ 40% of predicted normal for age, gender and height
- Subjects of child-bearing potential and who are sexually active must meet the contraception requirements

Golisano Children's Hospital

Pediatric Pulmonary Lab

Huff Cough

Huff Coughing: Controlled Coughing Technique

(Also known as Directed Cough, Assisted Cough, and Forced Expiratory Technique-FET)

Coughing is the natural way of removing foreign substances from the lungs. In certain lung problems there is an excess production of mucus, which must be removed to maintain open airways and allow you to move air in and out of your lungs effectively. Retained mucus contributes to the risk of infection and increased mucus production. In addition, the mucus irritates nerve endings into the tracheobronchial tree and can cause frequent, involuntary coughing, which can make you very tired. Ineffective coughing, can also lead to airway compression (closing of the air tubes) making it so you cannot clear or get rid of the mucus. Huff cough is a gentle way of coughing, which attempts to speed up airflow while keeping the glottis (throat) open. To conserve energy and oxygen, you must practice and master the method of controlled "Huff Cough".

Huff Cough Instructions:

- Begin in a sitting position with your chin slightly forward
- Use your diaphragm (stomach muscle) to breathe in slowly (belly should go out when you take air in)
- Hold your breath for 2-3 seconds
- Blow air out quickly saying the word huff and **STOP** just before you feel the urge to cough (Do NOT cough)
- Repeat these steps again 2-3 times

Cough normally to get rid of the mucus

Adjusting therapy:

- The length and force of the breath will change depending on where the mucus is.
- A normal size breath clears the larger breathing tubes in your lungs
- A larger size breath clears the smaller more distant breathing tubes in your lungs.
- You should never Huff Cough to the point of exhaustion!
- Remember to keep your throat wide open when blowing out.
- If you hear a "wheezing sound", blow out more softly
- Cough when you feel the mucus has collected in your larger breathing tubes.

Practice, practice, practice!

Huff coughing can be done anywhere at anytime…you control it……

Don't let your cough control you!

Fun Page!!

Show us your creative skills!
Enter our first ever coloring contest!

Pictures are posted on our webpage for you to download and print as many copies as you need. Only one may be submitted. We have 3 age groups and 1 winner for each group. Age groups are; 5 and under, 6 to 11, 12 and up. Prizes may include: Free meal at McDonalds, Free admittance to Kanga Play Center, Free admittance to The Strong Museum of National Play, or an iTunes gift card. CF center staff will judge the entries and notify winners on September 7th. You may drop off your entry at a clinic visit or mail to:

Melissa Berry, RD
Pediatric Pulmonary
601 Elmwood Avenue, Box 667
Rochester, NY 14642

Find the coloring pages at: http://www.urmc.rochester.edu/childrens-hospital/pulmonology/cystic-fibrosis/resources.cfm

If you do not have access to a computer or printer, call 585-275-1457 to request a coloring sheet and we will gladly mail one to you! Make sure you write your full name and age on the back of your entry.

"Prescription" for a hot summer day: Homemade Ice Cream in a Bag!

What you'll need:

1 tablespoon sugar
1/2 cup milk or half & half
1/4 tsp vanilla
6 tablespoons rock salt
1 pint size plastic food storage bag
1 gallon size plastic food storage bag
ice cubes

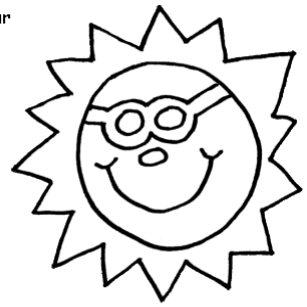
How to make it:

1. Fill the large bag half full of ice, and add the rock salt. Seal the bag.
2. Put milk, vanilla, and sugar into the small bag, and seal it.
3. Place the small bag inside the large one, and seal it again carefully.
4. Shake until the mixture is ice cream, which takes about 5 minutes.
5. Wipe off the top of the small bag, then open it carefully. Enjoy!

Tip: A 1/2 cup milk will make about 1 scoop of ice cream, so double the recipe if you want more. But don't increase the proportions more that that -- a large amount might be too big for kids to pick-up because the ice itself is heavy.

Unscramble the letters to find the words in our

Summertime Anagram



Hidden Word (circled letters): _____

a f n _____

a a b b e l l s _____

b e e f i r s _____

d g h o o t _____

h o r s s t _____

a c g i m n p _____

e f l o r w _____

a d e g n r _____

g i n s w _____

n s u _____

http://www.dltk-holidays.com/

University of Rochester
Medical Center
Cystic Fibrosis Center

601 Elmwood Ave
Box 667
Rochester, NY 14642
Pediatric Phone: 585-275-2464
Pediatric Fax: 585-2758706

Adult Phone: 585-654-5432

WWW.URMC.ROCHESTER.EDU/
CHILDRENS-HOSPITAL/
PULMONOLOGY/CYSTIC-FIBROSIS

CF Cycle for Life: Wheels in Motion to Cure CF

Saturday, August 20

Join us for the FIRST annual Finger Lakes Aptalis CF Cycle For Life®! The route begins and ends in historic Geneseo, NY and offers beautiful views of the Finger Lake's rolling hills. Ride at your own pace – this event is not a race, but a day to enjoy the beautiful countryside and celebrate your efforts to improve the lives of people living with Cystic Fibrosis.

For more information go to www.cff.org/Chapters/rochester

This event is for ages 18 and older only.

Are YOU the next CF Chef???

The CF Care Forward program is sponsoring a CF Chef Challenge Recipe Contest!!! Keep your eye on www.Chef4CF.com for details—more info will be posted by the end of June!