August 2012

The CF Breeze

"Wisdom & inspiration for CF families"

Clinics Visits

What to expect and why they are important!

After several years of 4 'uneventful' appointments per year and increased co-pay, my husband and I were beginning to question the value of bringing our daughter Kyra to so many appointments at CRCCS. However, we were recently reminded of the importance! At Kyra's quarterly appointment in July, the results of the routine throat culture indicated that she had a lung infection. Since the infection was caught early, it was still mild and Kyra had not developed any symptoms. Without that regularly scheduled appointment and throat culture, the lung infection would have gone undetected and likely become worse. After the diagnosis, Kyra started on the appropriate treatment ("Tobi torture" as she calls it) and we are hopeful that the infection will go away quickly. This was a 'good' reminder to us of the importance of the four yearly appointments. We certainly won't be missing any!

Amy S.
Mother of Kyra, Age 8

The CF Foundation is a leader in the efforts to advance and promote specialized care that improves and extends the lives of people with CF by detecting infections early. To ensure this continues, the Foundation provides its accredited care centers with up-to-date care guidelines based on the latest research. CF Foundation guidelines include a minimum of **four quarterly visits per year** for all patients with the diagnosis of cystic fibrosis. The following is an example of what the four visits would include:

- **Annual Blood Work and Chest X-Ray Appointment**: Typically, this will be the first appointment of the year. The patient will have throat or sputum culture, blood work taken, chest x-rays, PFTs (age 5 and over) and an office visit with the primary pulmonary provider. The blood work evaluates liver function, vitamin levels, blood count, and nutritional status. In addition, an oral glucose tolerance test is included for all patients age 8 and over.
- **Annual CF Team Appointment:** Typically, this is the appointment closest to the patients birthday. The patient will have throat or sputum culture, PFTs (age 5 and over) and meet with multiple members of the CF team as well as their primary pulmonary provider. The intent is to provide educational updates and get questions answered. When your child is between the ages of 6-8 we will coordinate a visit with the endocrinologist at the annual clinic to review the potential complications of cystic fibrosis related to the endocrine system.
- **Other two appoints throughout the year:** Typically, at these appointments the patient will have a throat or sputum culture, PFTs (age 5 and over) and an office visit with the primary pulmonary provider.

As a reminder, volunteer services is available during the annual CF team appointment to watch the children if families want to talk to any CF Team members alone without distractions

Upcoming Events

August 27 - CF Parent Support Meeting
Children's Minneapolis

September 22 - Aptalis CF Cycle for Life®
Lake Minnetonka

September 28 - Seven Courses to a Cure
McNamara Alumni Center, Minneapolis

September 24 - CF Parent Support Meeting
Children's Minneapolis

For more information, visit:
www.cff.org/Chapters/minnesota

The CF Breeze will be distributed electronically for patients and families of Children's Hospital and CRCCS. Please contact Mahrya Johnson at Mahrya.Johnson@childrensmn.org to be added to the distribution list.

The ABC's of Back to School!

Allergies: Let the school nurse know if your child has any allergies.

Bathroom: Let your child's teacher know that your child should have extended bathroom privileges.

Care Team: Remember that your child has a care team at school and in clinic. Don't be afraid to ask for help as you embark on this educational journey. You are not alone.

Diet: Remember to pack lunches and bring snacks that are high in calories and fat!

Enzymes: Determine who will dispense the enzymes when your child is at school.

Friends: What would you and your child like communicated to their classmates?

Germs: Pack antibacterial lotion in your child's backpack and encourage regular use throughout the school day.

Hydration: Make sure your child has access to plenty of fluids. This is especially important on hot and humid days.

Individual Health Plan: How will the school staff know what your child needs to remain healthy?

Juggle: Prepare yourself, and your child, to juggle a schedule balanced between treatments, school, family and fun. What might a typical school day look like?

Knowledge: Share what you know with the school staff and all those working with your child. You are their best advocate!

Loop of communication: How will you communicate with the school? Is there a plan in place for infection control?

Medications: The school should have a list of your child's medication on file.

Nurses Office: The nurse is your child's friend! Call the nurse prior to school starting to let them know your child needs.

Organization: Visit www.cff.org and connect with your local chapters for support, educational resources, and updates.

Parent Support Group: The support group is a wonderful opportunity to talk with other parents about managing school.

Quarantine: Don't be afraid to keep your healthy child with CF home when others at school are sick.

Respiratory Care: Remember to keep your child's lungs as healthy as possible by sticking with their respiratory therapies.

Separation: In the event that more than one child with CF attends your school, keep them 3 feet apart at all times.

Treatments: You may need to modify your child's current treatment schedule as they transition back to school and are engaged in more activities.

Unique: Although there are a lot of common characteristics among children with CF, always remember that your child is unique and may have needs that differ from others affected by CF. Speak up and share!

Vitamins, Vitamins, Vitamins! They help provide your child with all the nutrients their brain will need to soak up all the new information they'll be learning!

Water breaks: Inform your child's teacher that water breaks are important!

e**X**pect great things: Your child's post-secondary options are endless. Help them to set, obtain, and live their dreams each and every day.

Yes you can!: Managing CF care and school is a tough job, but with a team effort...you can do it ;)

ZZZZZ's: Sleep is so important! Make sure your child is getting enough sleep at night so they are ready to learn!

Salt, Fluids and Cystic Fibrosis

Why Hydration is important

Most of the time, people with CF regulate their salt and fluid levels by adding salt to their food, choosing salty foods, and drinking to match thirst. Under "normal" conditions, this balance of salt, fluids and hydration levels in the body is maintained. There are many special circumstances, however, when more salt and fluids are needed to keep a person with CF well hydrated and felling well.

In infants and young children with CF small amounts of salt should be given throughout the day because human milk, formula, and baby foods are very low in salt. Guidelines recommend 1/8 tsp. daily for infants up to 6 months and ¼ tsp. daily for infants 6 months to 1 year. As the infant transitions to table foods, salt can be added to food, and many foods contain higher amounts of salt.

Anyone with CF can have an increased risk for salt depletion and/or dehydration with high levels of activity, fever, or hot, humid weather. Preventing these problems requires close monitoring. Children and teens are not always able to sense their increased needs for salt and fluids in strenuous conditions, so careful attention should be paid to the body's signals of dehydration.

Symptoms that may indicate dehydration and salt depletion include:

- Fatigue/exhaustion
- Confusion
- Weakness or loss of coordination
- Fever (increased body core temperature)
- Profuse sweating
- Muscle cramps
- Abdominal pain
- Vomiting

Extreme dehydration can turn into a heat stroke and requires immediate medical attention.

To prevent dehydration, adequate salt and fluids are necessary throughout the day, and especially so during strenuous activity or when sweating is likely. People with CF should add salt throughout the day. Foods that are salty include: pretzels, potato chips, crackers, popcorn, canned soups, pizza, processed cheese, soy sauce, and lunchmeats. Salt should be used generously when cooking and sprinkled on foods once they are prepared.

Fluids should also be increased before, during and after activity. Cold, non-carbonated beverages are recommended for use with activity in heat. For children and adults with CF who are participating in sporting events, eating salty foods might not be possible, and standard sports drinks may help boost the salt level but may not be enough.

Salt depletion and dehydration are serious health concerns for people with CF. Eating enough salty foods, adding salt to food and drinking plenty of liquids can prevent dehydration. Sickness, hot weather and strenuous activity are all situations that require extra attention. Please call your CF Team if you have any concerns about dehydration or need more specific guidelines.

(Parts of this article were adapted from Children's Hospital of Wisconsin CF Program Handout)

CF Foundation Research Updates

If you would like to learn more about the CF Foundation research activities, please visit their website at http://www.cff.org/research/.

Nutrition Basics

Research has shown there is a strong correlation between nutrition and lung function in CF patients. Ensuring good nutrition is one of the main goals for patients and families. Nutritional basics for CF includes:

- **Diet.** People with CF need a healthy, balanced diet of all food groups that is very high in calories. It should also be high in fat, protein and salt.
- **Enzymes.** Most people with CF take enzymes to help them break down foods in order to effectively absorb nutrients.
- **Vitamins.** Vitamins A, D, E and K are fat-soluble vitamins. Since digesting fats is a problem for most people with CF a higher amount of vitamins is required.
- **Salt.** Salt is essential for people with CF for muscle function, nerve cell health and to improve the flow of fluid in cells. Since patients are losing higher than normal levels of it when they sweat, replacing this loss is very vital

Featured Recipe

Macaroni and Cheese Dinner

This classic is now packed with more calories and fat. The creamy cheese flavor is sure to win over even the pickiest eater.

Ingredients:

6 c. water
1 (7.25 oz.) package of macaroni and cheese dinner
¼ c. butter
¼ c. heavy whipping cream
2 tbsp. skim milk powder
½ c. cheddar cheese, shredded

Directions:

1. *Boil the water in a medium pan.*
2. *Stir in the macaroni.*
3. *Boil for 7 to 10 minutes, stirring occasionally.*
4. *Drain the macaroni and return it to the pan.*
5. *Add butter, whipping cream, skim milk powder, cheese, and cheese sauce mix (from the macaroni and cheese dinner package).*
6. *Reduce heat to low and mix well until the cheese has melted.*

Recipe from the following website: http://kidshealth.org/parent/recipes/cf_recipes/about_cf_recipes.html

Medication 101

Enzymes

Current FDA approved enzymes for CF include Creon®, Zenpep, Pancreaze™ *(formerly Pancrease)****, Ultresa*** *(formerly Ultrase),* ***Viokace*** *(formerly Viokase)* ***and Pertzye*** *(formerly Pancrecarb).*

Purpose: Pancreatic enzyme products (PEPs) contain the active ingredient pancrelipase, a mixture of the digestive enzymes amylase, lipase, and protease. These digestive enzymes are used to improve food digestion in patients with CF.

Enzymes are used to improve digestion of foods and prevent frequent, fatty, foul-smelling bowel movements in people who have a condition that affects the pancreas such as cystic fibrosis. Pancreatic enzyme products act in place of the natural enzymes normally made by the pancreas. It works by breaking down fats, proteins, and starches from food into smaller substances that can be absorbed from the intestine. This allows the body to use these substances for energy and prevents them from being passed as frequent, fatty bowel movements.

Potential Side Effects: may include diarrhea, constipation, upset stomach, vomiting, stomach cramps, bloating and gas.