May 2012

The CF Breeze

"Wisdom & inspiration for CF families"

Welcome to the First Edition of the CF Breeze!

On behalf of the CF Family Council, welcome to the first edition of the CF Breeze. The CF Breeze is a quarterly newsletter for families affected by CF that will highlight patients and Children's Hospital staff, provide nutritional information and recipes, research updates, and treatment-related material. The CF Family Council will be accountable for publishing this newsletter.

Upcoming Events

May 5

Great Strides Walk

Minneapolis, Lake Calhoun North Beach

May 20

Great Strides Walk

St. Paul, State Capital Building

June 25

CF Parent Support Meeting

Children's Minneapolis

July 25

CF Parent Support Meeting

Children's Minneapolis

July 16

65 Roses Golf Classic

Prior Lake, Legends Golf Club

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

The CF Family Council at Children's Hospital and Clinics of Minnesota was formed in May, 2011 as a result of CF parent requests and best practice recommendations from the CF Foundation.

The CF Family Council exists to foster partnerships between children with Cystic Fibrosis, their families, their care team and the CF Foundation. These partnerships will enhance both the patient and family experience with all facets of care, as well as, improve health outcomes of the children through empowerment, communication and education.

The Council is currently made up of:

- 5 Parents - individuals whose children are currently patients with CF at Children's Respiratory & Critical Care Specialists (CRCCS)
- 3 Children's Hospital and CRCCS care providers, Mary Sachs, RN, CNP, Sandy Landvik, RN, and Lacie Johnson, LICSW
- Children's Hospitals Family Coordinator, Tessa Billman.

Since its inception in 2011, the Council's accomplishments include:

- Development of a travel brochure – providing insight and tips for families traveling with children with CF
- Creation of a travel template form – for CRCCS staff to use in assisting traveling patients
- Initiation of CRCCS Patient/Family listserve (e-mail distribution list) – to improve ease in disseminating information from the clinic to patients and families
- Input/feedback to CRCCS in regards to CF Patient Annual Visit - to improve all aspects of the visit, from scheduling to the team visit.

The Council would like to have members rotate on/off on a regular basis, approximately every 2 years. Please contact Sandy Landvik or Mary Sachs if you are interested in becoming a participant on the CF Family Council.

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.

Traveling with Cystic Fibrosis

Summer's (Almost) Here!

Don't let the fact that you or your child has Cystic Fibrosis (CF) stop your family from having a great vacation.

The CF Family Council has developed a brochure that can help you and/or your child prepare for and enjoy traveling. The most important thing to remember is that keeping up with the treatment routine will help your child maintain good health, and help your family enjoy your vacation to the fullest extent.

When traveling, plan your vacation and research your destination in advance. In addition, consider the following:

- Medicines and equipment needed while you are gone
- Documentation needed to get to your destination (CF travel form & care summary available from the clinic)
- Travel time; recognize limits to avoid becoming overtired and sick
- Tolerance for altitude, heat, activity, and food
- General and/or medical travel insurance, and refundable or flexible tickets
- Vaccination status – helpful information available through the Center for Disease Control at http://wwwnc.cdc.gov/travel/.

Talk to your health care provider if you have any concerns about your travel plans and/or current health status.

Prior to leaving on vacation, it is important to obtain the necessary health-related travel documents, which may include:

- travel form from your CF clinic with information such as a list of your medicine, food and drink requirements, any equipment needed (for air travel), and clinic contact information
- a simple summary of your CF care from your clinic outlining your condition, usual antibiotic combinations for an exacerbation, and anything else that may be required to provide treatment

The traveling with Cystic Fibrosis brochure is now available at the Children's Respiratory & Critical Care Specialists (CRCCS) clinic.

Medical Research Updates

What is Kalydeco?

Kalydeco (kuh-LYE-deh-koh) is a pill taken orally twice a day for the treatment of CF in people ages 6 and over with the G551D mutation. It was approved by the FDA on January 31, 2012.

How does Kalydeco work?

In gating mutations like G551D, the defective protein in CF moves to its proper place at the cell surface but does not function correctly. The CFTR protein instead acts like a locked gate, preventing the flow of salt and fluid in and out of the cell. Kalydeco acts to unlock that gate.

During clinical trials Kalydeco was found to help improve lung function and lower sweat chloride levels. It also helps patients gain weight and improve overall quality of life. These are all key indicators the drug is working.

Will Kalydeco work for other CF mutations?

It is hoped that future clinical trials will demonstrate that people with other CF mutations similar to G551D will also respond to Kalydeco. There are presently 4% of people with CF that have the G551D mutation. Patients who are eligible have been notified.

Medical Research

What a difference you are making in the lives of Patients and Families….

Children's CF Research program has grown significantly over the past 5 years. It is our mission to not only improve the quality of care we provide, but to improve the overall quality of life for the patients we serve.

Thank you to all the patients and families who have donated their time and energy participating in CF related clinical trials. You are the key to finding a cure. Without your help, research cannot move forward. We need your continued participation to move promising therapies from the testing phase to the people who need it most.

Studies Open for Enrollment (Participants must meet specific inclusion/exclusion criteria to qualify)

- Evaluation of Sleep in Children and Adolescents with Cystic Fibrosis
- Quality Improvement in Genetic Counseling Following a False Positive Newborn Screen Result for Cystic Fibrosis: Assessment of Parental Knowledge with the Use of an Educational Video
- (Twin/Sibling) Genetic modifiers system of cystic fibrosis study
- Genetic modifiers system of cystic fibrosis liver disease

Studies Closed for Enrollment

- Evaluating the effects of yoga on children with cystic fibrosis: Pain, sleep, anxiety and depression
- (CSREP) Physical Therapy study cystic fibrosis core strengthening and respiratory exercise program
- The EPIC observational study: Longitudinal assessment of risk factors for and impact of *Pseudomonas aeruginosa* acquisition and early anti-pseudomonal treatment in children with cystic fibrosis (Protocol #EPIC-002).
- The incidence of pain, other physical symptoms, and depression/anxiety in pediatric patients with cystic fibrosis: Impact on overall quality of life, and eating disorders-body image

Other Research

- Evaluating the Effects that *Bordetella bronchiseptica* has on the CF Patient Population
- Incidence of Cystic Fibrosis and CFTR Mutation Distribution in the Minnesota Newborn Screen Population
- MRSA Characteristics in a Cystic Fibrosis Pediatric Population

University of Minnesota Studies

- Prediction by Ultrasound of the Risk of Hepatic Cirrhosis in Cystic Fibrosis (PUSH)

To learn more about the Cystic Fibrosis Research Program, contact Mahrya Johnson at (612) 813-6384
Mahrya.Johnson@childrensmn.org

Staffing Updates
Lisa Read, Clinical Research Coordinator, has taken on a new role at Children's and will be missed as part of the CF team. Andrea Gruber has been hired to replace Lisa and will be stopping by at clinic visits to introduce herself.

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

An important factor for staying healthy is good nutrition. Thick mucus often gets in the way of proper digestion, causing malabsorption. This problem is treated with pancreatic enzyme supplements, vitamins and a high-calorie, high-fat diet.

A high-calorie, high-fat diet is vital for normal growth and development in children with CF, and offers adults a way to maintain optimal health. The dietitians at CF Foundation-accredited care centers work with patients and their families to map out the best diet for each person.

Nutrition and general lung health are closely linked. People with CF may need extra calories to compensate for the malabsorption of nutrients. These extra calories also help to meet the greater energy needed for breathing. In fact, for children with CF, extra fat calories are good for fueling normal growth and development.

Below is a recipe for a high-calorie, high-fat and gluten-free snack.

Featured Recipe

Peanut Butter Bacon Cookies

1 cup creamy peanut butter
½ cup granulated sugar
½ cup brown sugar
1 egg
1 tsp. baking soda
6 slices bacon, cooked, cooled and diced

Directions:

1. In a skillet, fry bacon and set aside
2. Pre-heat oven to 350 degrees
3. Grease cookie sheet with butter, set aside
4. In a mixer, combine peanut butter and sugars for about 2 minutes until well blended
5. Add egg and baking soda and mix for another 2 minutes
6. Fold in cooked bacon (cooled and diced)
7. Roll into walnut-sized balls, coat in granulated sugar, flatten with a fork and place on a cookie sheet
8. Bake for 10 minutes until lightly browned
9. Cool on the cookie sheet for 5 minutes, then enjoy!

Found in Iowa Alumni Magazine – reprinted from Joy the Baker. www.joythebaker.com

Medication 101

Albuterol

Purpose: Albuterol is in a class of medications called bronchodilators. This medicine opens the airways by relaxing the muscles around the airways, making the air passages to the lungs larger. This, in turn makes breathing easier. Albuterol is used to prevent and treat wheezing, difficulty breathing and chest tightness caused by lung disease, such as asthma. Albuterol allows for better air flow and easier airway clearance, and is routinely used during airway clearance treatments to help clear mucus from the air passages.

Potential Side Effects: may include a fast heartbeat, shaky hands, headache, or jitteriness. These side effects should decrease as the body gets used to the medicine.

Pulmozyme

Purpose: Pulmozyme (dornase alpha) is a mucolytic, which helps break up mucus in the lungs and makes the mucus thinner. DNA in white blood cells accumulate in your lungs in response to an infection. After white blood cells fight the bacteria, they leave behind extracellular DNA that can make mucus more thick and sticky. Cutting up the extracellular DNA can help thin mucus. Pulmozyme breaks down or cuts through the DNA of white blood cells, making the mucus thinner. When the mucus is thinner, it is then easier to loosen, mobilize, and cough out.

Potential Side Effects: may include voice changes (hoarseness), sore throat, chest pain, red watery eyes, fever, headache, tiredness, increased cough. These side effects are usually mild and temporary.