

February 2014

The CF



Breeze

"CF wisdom & inspiration for CF families"

NEW! Beads for Breath Program



We are pleased to announce the rollout of a pilot program called Beads for Breath!

We know living with CF is time consuming and can have an emotional impact on both children and their families. The goal of this program is to provide education, support, and coping strategies to children living with CF around their treatment plans by earning beads for various goals and procedures.

Throughout history, Beads have symbolized courage and strength. Arts in medicine programs like Beads of Courage (oncology) and Heart Beads have been shown to have a positive emotional impact for parents of chronically ill children, and the patients themselves. The beads help create a narrative for the child and family to communicate about their disease and the medical treatment they have to endure. With the life expectancy for cystic fibrosis continually increasing, it's imperative to create and implement similar programs that help improve psychological adjustment and understanding of their disease to promote long term health management.

Who's Eligible?

All children with CF between the ages of 5-10 years of age on their annual visit.

How Do I Sign My Child Up?

Your Social Worker, Lacie Johnson, will meet with you on your annual clinic visit and will review program details. If you are interested in the program your child will begin earning beads that day!

How Does My Child Earn Beads?

There are 23 different ways your child can earn beads. Examples include: attending quarterly clinic appointments, formal PFT's, and being hospitalized.

Who Distributes The Beads?

Various staff members will be distributing beads. A Bead Handout will be provided at the time of enrollment.

If you have additional questions, please contact Lacie Johnson at 612-813-6839.

Upcoming Events

March 24

7:00 pm to 9:00 pm

CF Parent Support Meeting

Children's Minneapolis

March 28

Brewer's Ball

Minneapolis, TCF Bank Stadium, DQ Club Room

April 28

7:00 pm to 9:00 pm

CF Parent Support Meeting

Children's Minneapolis

May 3

Great Strides Walk

Minneapolis, Lake Calhoun North Beach

May 18

Great Strides Walk

St. Paul, State Capital Building

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.

To be added to the distribution list or to update your e-mail address, please complete a listserv form available in the lobby of CRCCS or by contacting Mary Sachs or Sandy Landvik.



New Infection Control Guidelines for people living with CF

The CF Foundation recently updated its infection control policy in the hope of maintaining the “health and safety of people with CF wherever they gather”. This applies to whether that location is in a clinic office space, at a school or CF Foundation event. The goal of this updated policy is to limit the risk of cross-infection between people with CF and is based on medical evidence that supports that all people with CF could have germs that might be spread to others with CF. The CF Foundation’s 10 page document provided to all CF Centers discusses these specific recommendations and the supporting references and rationale for putting these guidelines into practice.



A summary of the specific recommendations that are a part of this document are listed below. As with all recommendations and guidelines, it is important to remember that you may not be able to prevent all infections, but you can reduce your risk.

Recommendations:

1. Only one person with CF may be present at a CF Foundation-sponsored event that is indoors.
2. At CF Foundation sponsored outdoor events, people with CF need to be 6 feet apart.
3. If there is a past or present history of a confirmed positive sputum culture for B. cepacia, this person may NOT attend any CF sponsored event.
4. EVERYONE (regardless of having CF or not) should clean their hands using soap and water or an alcohol based hand gel that is at least 60% alcohol. Hands should be cleaned after coughing, sneezing, blowing the nose, before eating and after going to the rest room.
5. All people with CF should avoid any activities with other people with CF because of the risk of germ transmission. These include: shaking hands, hugging, riding in the car together, sharing hotel accommodations, exercise classes or sharing any cups or utensils.
6. Information about whether a person has CF or what germs that they have in their lungs is to be maintained as confidential unless the family wishes to have this information known.
7. Patients with CF must wear a mask in the waiting room and throughout the health facility except in their own exam room (clinic or hospital room). Infants and young toddlers may have a blanket placed gently over their face.
8. Staff in the clinic and hospital room wear will gown and gloves when seeing the patient in their exam or hospital room.
9. Schools should avoid common lunch room times, classrooms and recess times.
10. Children or adults with CF should avoid hot tubs, whirl pool tubs, and construction sites. They should also avoid cleaning stalls, pens or coops.
11. Patient exam rooms should be cleaned between patients as well as stethoscopes.
12. People with CF who live in the same household should not share utensils, tooth brushes, or respiratory equipment. Whenever possible, they should also perform airway clearance with only one person with CF in the room at the time of treatment.



2012 CF Outcomes

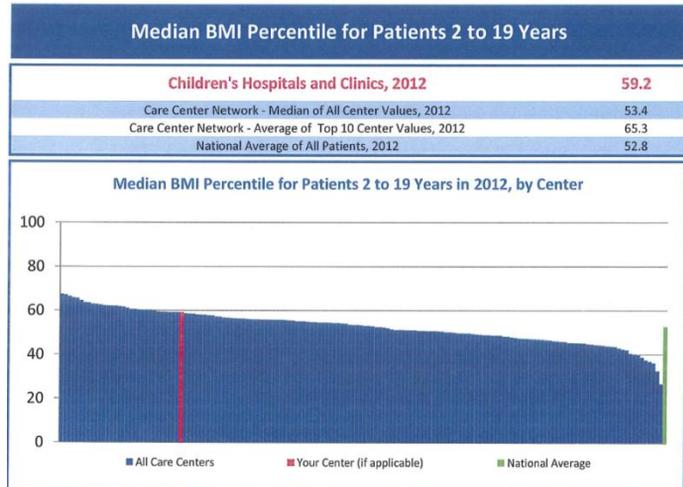
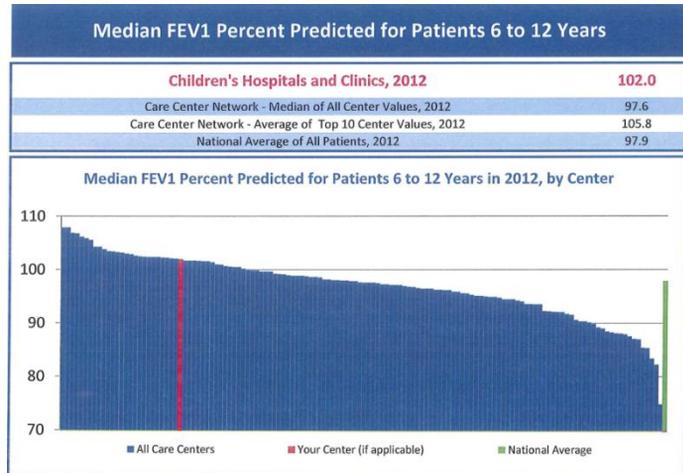
Children's Hospitals & Clinics of MN

Each year our CRCCS/Children's Hospital CF Care Center receives a report on measured outcomes from the CF Foundation Registry. This is a result of you / your child voluntarily participating in the data entry. Two slides from that report are to the right- they represent the 2012 pulmonary and nutrition outcomes for our Center.

The FEV1 slide shows the median lung function of patients in our center for 6-17 year olds to be in the top ten compared with all the centers in the country. Normal lung function in the general 6-17 year old population is between 80-120%. Our patients with cystic fibrosis have a median lung function of 102%!

The BMI slide shows that the patients at our center have a median BMI at the 59th percentile. The goal we are aiming for is the 50th percentile. Research has shown a direct correlation between BMI (body mass index) and lung function. "The better the BMI the better the lung function."

These two slides show how well CRCCS/Children's Hospital CF Care Center performs compared to the rest of the CF centers in the country. It is a tribute to the hard work you do to improve the outcomes of this disease. More information on Children's Center data is located on the CFF web site at www.cff.org.



Becoming a HopeKid

HopeKids provides ongoing free events & activities and a powerful, unique support community for families who have a child (0-18) with cancer or some other life-threatening medical condition (e.g. Cystic Fibrosis). HopeKids surround these remarkable children and their families with the message that hope can be a powerful medicine.

How to enroll:

- Submit the HopeKids application. <http://www.hopekids.org/become-a-hopekid/>
- Medical eligibility will be reviewed with the treating physician.
- If it is determined that your child is eligible, you will then receive an email with instructions on how to complete your registration.
- Once you complete the registration process, you will begin receiving invitations by email to our free events each month.

There is never a cost for joining HopeKids and all events are free, however there may be some expenses you choose to incur (i.e. food at a ballgame, parking ramp at a venue, souvenirs at a concert). The majority of the ongoing program of events centered around the greater metropolitan area of Minneapolis/St.Paul.

** Please note: there may be other children with CF that attend these events. HopeKids attempts to separate children with CF when possible.*



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)

- Early MRSA therapy in CF – culture based vs. observant therapy (treat or observe) for patients ages 4 – 45 years old. (STAR-too-**ST**aph **A**ureus **R**esistance – treat **or** observe).
- **B**aby **O**bservational and **N**utritional **S**tudy (BONUS) for patients less than 3.5 months old.
- A Long-Term Prospective Observational Safety Study of the Incidence of and Risk Factors for Fibrosing Colonopathy in US Patients with Cystic Fibrosis Treated with Pancreatic Enzyme Replacement Therapy: A Harmonized Protocol Across Sponsors.
- Clinical Outcomes in Pediatric Patients with CFTR-Related Metabolic Syndrome (CRMS) for patients with CF or CRMS and were diagnosed by newborn screening between 2006-2015. The CF Foundation has developed Practice Guidelines for managing patients with CRMS, but we would like to improve our understanding of CRMS.

CF Foundation Research Updates

Have you had a chance to meet Emma Green yet? Emma is the main character in the book, "Emma Green: Science Superstar," about a young girl with CF who is offered an opportunity to participate in a clinical trial.

The story describes Emma's personal struggle with the decision about whether to become a study subject, as well as what happens once she does make up her mind to participate. Copies of Emma Green will be available in the lobby of CRCCS by the beginning of March.

An electronic version of the book is available on the CFF website.

- <http://www.cff.org/UploadedFiles/research/ClinicalResearch/Clinical-Trials-Booklet.pdf>

Link to Vertex Research Press Releases:

If you would like to learn more about the Vertex research activities, please visit their website at:

- <http://investors.vrtx.com/releasedetail.cfm?releaseid=743425>
- <http://investors.vrtx.com/releasedetail.cfm?releaseid=757597>



Research Contacts

To learn more about the Cystic Fibrosis Research contact:

- Mahrya Johnson: Office 612-813-6384; Email: mahrya.johnson@childrensmn.org
- Andrea Gruber: Office 612-813-6661; Email: andrea.gruber@childrensmn.org

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/>

CF Listserv Update

Please remember to submit email address changes by completing a listserv form and handing it to your clinic nurse. These forms can be found in the lobby of CRCCS, or by asking Mary Sachs or Sandy Landvik.



Featured Recipe

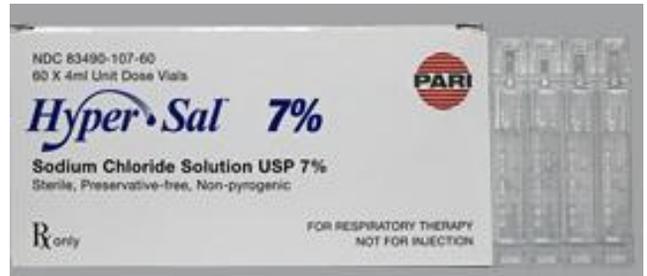
Reese's Chocolate Peanut Butter Rice Krispie Treats

Ingredients

- 6 cups Cocoa Krispies Cereal
- 16-18 Reese's Peanut Butter Cups, diced
- 5 Tbs butter or margarine
- 3 Tbs light Karo syrup
- 1 (10 ounce) package plus 2 heaping cups miniature marshmallows
- 1/3 cup peanut butter
- 1/2 cup chocolate chips

Instructions

- In a large bowl, combine cereal and diced Reeses. Set aside. Spray a 9 x 13 in. pan with cooking spray and set aside.
- In a large pot melt butter over low heat.
- Add karo syrup and marshmallows and stir continuously until mixture is smooth.
- Remove from heat and add cereal mixture into the pot and fold together just until cereal is evenly coated. (Peanut butter cup pieces will fall apart as you fold together your ingredients, so don't over mix this.)
- Press mixture into the sprayed 9 x 13 in. pan. Set aside.
- In a small microwave safe bowl, add peanut butter. Microwave for 30 second to a minute to melt. Set aside to let cool slightly.
- In a separate small microwave safe bowl, melt chocolate at 30 second intervals, stir between intervals, until smooth. Set aside to let cool slightly.
- When peanut butter is cool enough to handle, transfer into a small zip lock bag. Clip a small piece out of the corner and pipe onto rice krispie treats, zig-zagging back and forth.
- Repeat steps with chocolate, placing in a small zip lock bag, and piping over the peanut butter.
- Either let treats sit at room temperature to allow chocolate to set up, or you can pop the pan into the fridge for about 5 minutes to speed up the process. Cut and serve.
- Makes a 9 x 13 in. sheet of treats.



Medication 101

HYPERTONIC SALINE

What is Hypertonic Saline?

Hypertonic saline is a nebulized therapy used with children and adults with CF and approved by the CF Foundation as a daily preventative treatment. It is an extra salty (7%) sterile saline solution that enhances the clearance of mucus from the lungs by rehydrating the CF airway (which naturally lacks salt and water). It also helps stimulate cough which further improves clearance of airway mucus from the lungs.

How does it help in the treatment of CF?

Research studies demonstrate improved pulmonary function and fewer lung infections in patients with CF who use hypertonic saline twice daily. The usual age to start this medication is after age 6 years, but talk with your CF provider if you would like more information.

What are the Common Side Effects?

Hypertonic saline may irritate the airways and may also cause sore throat, chest tightness and cough. If any of these symptoms occur, a bronchodilator may be used first or at the same time if your CF clinic provider has prescribed the concentrated form of albuterol for you to use. Other reported side effects may be an unpleasant taste in their mouth after nebulizing.