

# CYSTIC FIBROSIS



Boston Children's Hospital



BRIGHAM AND  
WOMEN'S HOSPITAL

# News Wire

SPRING 2012

Dear members of our Cystic Fibrosis community:

It's been an exciting time since our last publication. Several new Fellows in training and Staff physicians have joined our ranks. The 2011 North American CF Meeting has come and gone, with a great deal of exciting new information about CF. Twenty-nine members of the CF Center Staff were at the meeting, and presented research findings, chaired sessions, and manned posters in the Scientific Sessions. In addition, we have seen the release by the FDA of the first gene-specific CF medication to help potentiate function of the protein made by the G551D defect. On all fronts, CF treatment is on the move.

As a CF Foundation Center, we work arm-in-arm with the Foundation and strive to closely follow the Foundation's Care Committee Guidelines. This means expanded interaction with many of our services, Nutrition and Physical Therapy (for two such examples). As we learn more about the impact of CF on our patients, the interventions necessary to keep patients in the best of health increase. This is how we improve on the survival and Quality of Life statistics.

While the new therapies are exciting, it should be obvious that we need to continue our vigilance and aggressive care. Some of the anticipated new therapies will be expected to help slow or stop the progression of CF disease, but will probably not retrieve better function from severely damaged organs. We keep working vigorously to take care of our patients until the disease has been beaten, and this will require hard work from everyone, especially our patients and their families, who are in the trenches every day with their own CF disease issues. Keep at it, and we will see you all at Great Strides in May!

*Henry L. Dorkin MD*

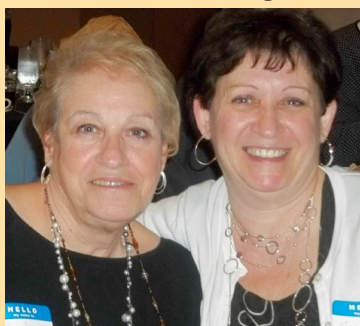
Cystic Fibrosis Center Director  
Boston Children's Hospital/Brigham and  
Women's CF Center

## CF Family Night

The Cystic Fibrosis Center at Boston Children's Hospital & Brigham and Women's Hospital CF Family Night was a great success! There were 140 family and staff in attendance. CF center leadership presented latest updates in the care and management of CF, research, and quality improvement. If you missed the evening look for the webcast on the Boston Children's CF center website.



*Amanda McGeachey, Olyn Andrade, Katherine Konaires, Ashish George, Robert Fowler and Georgina Garcia MD*



*MaryAnn Hebert and Jo Waite*



*Ahmet Uluer, DO*



*Soina Serrano and Evette Rodriquez*

## When my CF doctor recommends inhaled medications, which order should I do them in? When should I do airway clearance?

- Bronchodilator
  - Albuterol/xopenex
- Hypertonic Saline
- Pulmozyme
- Airway Clearance
  - CPT/VEST/Acapella
- Inhaled Antibiotic
  - TOBI/Cayston



*Michael Hebert*

## Pursuing your educational and career goals

### Leah Sands

Looking into the future is sometimes a challenge for someone with CF, especially because there can be a lot of uncertainty.

It may seem difficult to even think about going to college or working a regular job, particularly because a lot of our time must be spent on self-care and maintaining our health. Even for myself, I was never sure of what my future would hold, but I knew at a young age that I wanted to do my best to be a contributing member of society with the skills and talents I had.

It was during my junior and senior years of high school when I started to decide on my future college and career plans. I had always been interested in the medical field; it was a comfortable place for me to pursue a career after spending so many years as a patient. I applied for nursing school at a local college and was accepted. I also applied for several scholarships, both non-CF and CF-related ones. I was fortunate enough to have been granted two scholarships for my undergraduate degree - one from my college (for good grades and extracurricular activity involvement) and one through a CF-related pharmaceutical company. Both scholarships helped me a lot financially.

As I worked through the first couple years of college, I was able to live at home and commute to school, which was great. I scheduled my classes so that I could fit in my treatments and allow for sufficient study time. I was also able to maintain a part-time job at a medical laboratory doing administrative work. At the end of nursing school, I even took on a second part-time job so that I could move out of my parent's house and get my own apartment. I was thrilled that I was able to live an independent life! After five years of hard work and dedication, I graduated with my bachelor's in nursing in 2004.

However, shortly after nursing school, I realized that nursing wasn't quite my calling and that working in a hospital environment probably wasn't the best idea due to the chance for cross infection. I decided that medical administration was a better choice for me. I went back to school and got my medical transcription certificate in 2005 and was hired as a medical transcriptionist at a hospital.

It wasn't long afterwards that my partner and I moved to the East Coast for a change of pace. I landed a job in a doctor's office doing administrative work; the job was a great match for me. Also that year, we got married, and got pregnant with our first child.

Things were going so well with my job and our son that I got pregnant with our second child shortly after. By the time



### *Leah Sands and her sons*

our second son was born, my career was advancing nicely at the doctor's office. I was promoted three times during my employment there, ending up as their operations manager. Once I experienced a managerial role, I knew that management was my niche. But having a nursing degree wasn't really going to help me advance my career in management, so I decided to pursue avenues for my master's in business. I located several CF-related scholarships online. I was thrilled to see that one of the foundations offered a full-ride scholarship for an online program. I applied, and received a call that I was awarded one of the scholarships! What a great help that was financially, and even more motivation, for me to complete my master's.

So while my boys were still babies/toddlers, I worked through getting my master's while I continued working full-time at the doctor's office. It was hard managing my time, but I stayed focused and took everything day by day. Fortunately, I had a very understanding boss, so I was able to have some flexibility in my work day for doctor's appointments. After obtaining my MBA in 2010, I was able to land a fabulous job for a healthcare IT company. I now work out

of the clinical environment, but I am still involved in the healthcare industry, which is great.

Throughout the past several years, I have come to realize that I could achieve anything I put my mind to. Even when I felt that I couldn't do it, I kept focused and I knew the feeling of accomplishment would be worth it in the end. To be successful with school, work, family, and

health, I found that having a strict routine is key. I had my doubts about being able to maintain such a demanding lifestyle and career, but once I got out there and proved to my employers that I was a determined, hard-working person, there was never an issue with me needing time off for health concerns. I have also realized that there is a lot of help out there, particularly for people with CF. There are numerous college scholarships available, for both undergraduate and graduate degrees.

Being a working adult has really helped me maintain a sense of direction and control in my life. It's all about staying focused on what you want to achieve and making that your goal.

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*Leah Sands, 30 years old with CF*

*For more information on college scholarships, check out these web sites:*

*CF Care Forward Scholarship*  
[cfcareforwardscholarship.com/](http://cfcareforwardscholarship.com/)

*Boomer Esiason Foundation Scholarships*  
[cfscholarships.com/](http://cfscholarships.com/)

*Cystic Fibrosis Scholarship Foundation*  
[cfscholarship.org/](http://cfscholarship.org/)

*Elizabeth Nash Scholarship*  
[elizabethnashfoundation.org/](http://elizabethnashfoundation.org/)

# Screening and diagnosis of Cystic Fibrosis Related Diabetes (CFRD)\*

Cystic Fibrosis-Related Diabetes (CFRD) is a unique type of diabetes. It's not the same as diabetes in people without CF. The diagnosis and treatment are not exactly the same. CFRD is extremely common in people with CF especially as they get older. CFRD is found in 35 percent of adults aged 20 to 29.

## Causes of CFRD

There are two types of diabetes in the non-CF population - Type I diabetes (known as "insulin-dependent diabetes") and Type II diabetes (known as "non-insulin dependent diabetes"). tCFRD has some features of both types of diabetes. People with CF do not make enough insulin. This is a result of scarring in the pancreas. Insulin resistance is another reason people develop CFRD. Insulin resistance means your body does not use insulin normally.

## Symptoms of CFRD

Common symptoms, such as increased thirst and increased urination, are caused by high blood sugar levels (hyperglycemia). Other symptoms of CFRD are excessive fatigue, weight loss and unexplained decline in lung function.

## Screening and Diagnosis of CFRD

Many people with CFRD do not know they have it until they are tested for diabetes. Since many people with CF have no symptoms, this is the best way to find out if someone has CF-related diabetes.

The Cystic Fibrosis Foundation, American Diabetes Association and the Pediatric Endocrine Society put together guidelines with recommendations that our patients should be screened with an oral glucose tolerance test (OGTT) once a year after age 10.

## Treatment of CFRD

People with CFRD who receive treatment for diabetes often start to feel better, gain weight and improve their lung function. Insulin is the medication used to treat CFRD. It allows sugars and proteins to move from the blood into the body's cells. It is used for energy and to build muscle. Keeping blood glucose levels at a normal or near-normal level helps you gain weight, feel better and have more energy. It also lowers the risk of problems caused by diabetes.

We will begin annual screening for CFRD on all CF patients 10 years and older. We will be sending more information on scheduling your annual OGTT. Speak with your CF team at your next clinic visit for any questions or concerns.

\*Adapted from the Cystic Fibrosis Foundation 1/23/12

## Siblings

### Lynne Helfand SW, Isabel Bailey SW and Judy Bond SW

Parents frequently discuss feeling conflicted and stretched thin providing care for their child with CF and having enough time and energy for their other children.

They worry about meeting the needs of all of their children and struggle with the limited number of hours in a day and how many of those hours are devoted to chest PT, neb treatments, and clinic visits etc.

Healthy siblings are adjusting to having a brother or sister with CF just as parents are. Life changes for the entire family. Siblings report a variety of feelings and concerns about their brother or sister with CF. Some children experience guilt that they are not sick and their brother/sister is. Some siblings have 'magical thinking' and may wonder if a bad thought they had about their brother could have caused the CF. Some children may be anxious about becoming sick themselves and/or may wish they were sick too so that they could be the "center of attention."

For parents it's hard to give the attention that your healthy children may need while both managing all the care required for your child with CF and experiencing the understandable anxiety parents feel about their affected child. Healthy children sometimes can feel "invisible" which may contribute to their demanding more attention. Siblings might be encouraged to participate and help their brother or sister with their daily routines, by perhaps watching a video together during VEST treatments. The presence of a family member with a health issue can provide opportunities for increased empathy, responsibility and adaptability for everyone in the family.

Parents try to balance the needs of all of the children in the family and spending time with each child individually can help in nurturing a unique and special relationship with them. Siblings in addition need accurate information about their brother or sister who has CF and opportunities to talk about their concerns and questions with parents so that they feel heard and understood.

There are warning signs that may indicate that siblings may need some



*Lynne Helfand, SW, Isabel Bailey, SW and Judy Bond, SW*

extra attention or potentially some professional help. Do they seem:

- Anxious
- Depressed
- Withdrawn
- Angry
- Losing interest in friends or activities that once brought pleasure
- Doing poorly in school or pushing too hard to achieve
- Rebellious
- Blaming herself for her sibling's illness

Children all adapt differently when one member has a chronic health problem. Many factors can influence this process, including the course of the disease and the resources available to the family. While families with chronic health problems may struggle through difficult times it can be because of this relationship that siblings develop a sense of resilience, compassion and caring.

## Research updates

### Erin Leone Thakkallapalli, MPH, CCRC

In 2011, nearly 1,000 patients participated in CF research studies across the country. At Boston Children's Hospital, over 470 patients and their family members enrolled in observational studies and 46 subjects participated in interventional research studies. We want to take this opportunity to thank these patients and families for their involvement in CF research. These individuals participated in studies such as:

- Vertex VX-770 (ivacaftor) for Genotype G551D
- Gilead AZLI for Burkholderia
- PTC Ataluren (PTC124®)
- Mpex Aeroquin™
- Vertex VX-809

- Genetic Assay Validation
- iCARE

We hope to continue our center's contributions to CF research in the coming year. In 2012, Cystic Fibrosis Foundation Therapeutics is projecting that more than 1,530 subjects will need to enroll in research studies. We are currently enrolling subjects in the following studies:

- Gilead AZLI for Children with Chronic Pseudomonas (PALS)
- Gilead AZLI for Children with New Pseudomonas (ALPINE)
- G551D Observational (GOAL) Study
- Vitamin D Supplementation
- Acid Suppression Therapy

Additionally, we anticipate starting the following studies in the next several months:

- CF Fibrosing Colonopathy Observational Study
- FIRST Breastfeeding Study
- Vertex VX-770 (ivacaftor) for 2-5 year olds, Genotype R117H, and Non-G551D Gating Genotypes
- Several inhaled antibiotic studies

If you would like to participate in a study or learn more about CF research at Boston Children's, please speak to a member of our research team. Feel free to call us at (617) 355-6665 or ask for us during your next appointment.

## Summertime fun

### Kristen Leavitt RD, LDN and Julianna Bailey RD, LDN

With summer approaching we want to remind you of some important issues that often arise in patients with CF.

In hot, humid weather or in periods of exercise, people with CF are at risk of consuming too little salt and becoming dehydrated. All people tend to lose fluid and salt when they sweat and exercise, but people with CF can lose more. It is important to make sure that your child's fluid and electrolytes are replaced during this time.

Here are some ways to help your child get enough fluids and electrolytes during physical activity:

Make sure your child drinks plenty of water and sports drinks (Such as Gatorade® and Powerade®) to replace electrolytes lost in sweat during exercise.

Drink water before physical activity to help prevent dehydration (three to six ounces for children under 90 pounds and 6 to 12 ounces for children who are 90 pounds or more).

Once exercise begins, people with CF should drink six to 12 ounces of fluid every 20 to 30 minutes, even if they do not feel thirsty.

With heavy sweating, add an extra 1/8 teaspoon of salt to 12 ounces of a sports drink or to eight ounces of water.

Avoid carbonated drinks, such as sodas, and drinks with caffeine, during exercise.

If your child is not drinking enough fluids, you may notice:

- Dry mouth
- Fatigue and weakness
- Fever
- Heavy sweating
- Muscle cramps
- Abdominal pain and/or vomiting
- Changes in urine (darker and strong smell) or your child does not need to urinate as often.

If your child develops any of these signs, have him or her stop the activity, move into the shade or a cool place, and drink fluids right away. If the symptoms continue, you may need to get medical help.

### What are the best sources of salt?

Table salt is the best source of sodium chloride, along with foods that are processed with salt, like bacon and pickles. Fresh foods such as meats, chicken, fish, fruits, vegetables, rice and pasta have very little salt. But, these foods are high in salt when they are processed into canned and boxed soups, vegetables and pastas, and canned, boxed and frozen dinners. Many food manufacturers are lowering the salt in their products, so it is important to read labels and use the saltshaker to add extra salt. Food labels tell you how much sodium is in food; foods that are higher in sodium are higher in salt.

### Food Sodium (mg)

Ramen, Cup Noodles (1 package)	1430
Salt (1/4 teaspoon)	580
Dill Pickle (1 medium)	570
Canned soup (1 cup)	480
Hard pretzels (1 ounce)	385
Pedialyte® (1 cup)	245
Cheerios® (1 cup)	204
Gatorade Endurance® (1 cup)	200
Bacon (1 slice)	185
Potato chips (1 ounce)	180
Canned carrots (1/2 cup)	177
Gatorade® (1 cup)	110
Whole milk (1 cup)	98

## Hearing loss

Patients with cystic fibrosis may be at increased risk for hearing loss. Although the exact cause has not been identified, several routine CF therapies, particularly certain antibiotics, may lead to some damage to sensory hair cells in the inner ear, which may cause decline in the ability to hear. The Department of Otolaryngology and Communication Enhancement and the Cystic Fibrosis Center are working together to investigate specific factors contributing to hearing loss.

It is the goal of the CF center that patients with CF have an annual hearing test. Once a patient is identified as having a decrease in hearing, they will be seen more regularly by otolaryngology and audiology in order to keep careful record of any progression in hearing loss.

## Educational day for new families

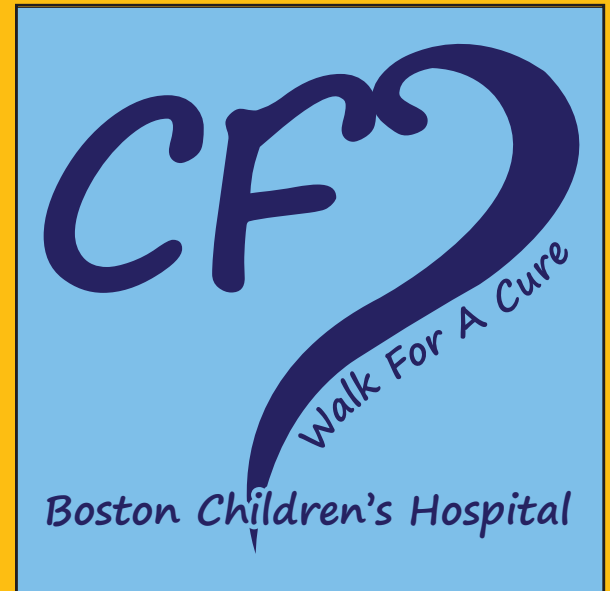
The CF Center hosted an educational day for new CF families. It was an informative day presented by the pediatric multidisciplinary team. Thank you to all who participated!

## Events

On June 17th, 2012 the Adult CF Program at Boston Children's Hospital and Brigham & Women's Hospital is hosting an educational event, *The 'Power of Two in You: Staying healthy, living with CF and lung transplant.*

Our event will touch on three major themes of adult disease management: managing your busy schedule around recommended chronic pulmonary therapies (including caregiver support); health maintenance with a focus on exercise; and organ transplant awareness/lung transplant. The main attraction for this event will be the powerful documentary, 'The Power of Two', which features two sisters of Japanese descent with CF and their journey involving lung transplant. The movie will be preceded by breakout groups for those interested to discuss a specific theme more in depth, followed by the entire audience and patients/others at home (via web streaming) having an open discussion. We intend to embed the movie and discussion into a Facebook group, so that patients and families, who are unable to attend in person, may still participate in our community experience. Furthermore, we will utilize methods for interaction with our home audience via Facebook, Twitter, and other media tools. We will be asking members of the CF community whether they wish to display their art in a small gallery space and we also hope to hold a small book fair. Our event will be held at Boston's Wheelock Family Theatre. We continue to look into other ways we might enhance your experience interacting with our event, so please stay tuned to updates and changes to our plans in the coming weeks. The tentative start time on June 17th is 1:30 p.m.

Thank you to Bianca Augeri, the winner of the CF Center Great Strides tee shirt contest



[www.ThePowerOfTwoMovie.com](http://www.ThePowerOfTwoMovie.com)  
*A story of twin sisters, two cultures, and two new chances at life—Directed and produced by Marc Smolowitz*

We follow Cystic Fibrosis Foundation Infection Control Guidelines. Because of the potential risk posed to the patient spread of bacteria, we regret that we are unable to invite patients with CF to attend this event. In light of this precaution, we encourage all patients to send family and friend representatives.

We invite patients and families unable to attend the event to view our presentation live via webcast starting at 2PM on June 17th. To do so, we will provide an appropriate link at a future date, please stay tuned. During the webcast, you will be able to view all presentations and submit questions online in real-time. Please note that you should have no concern regarding transmission of bacteria between parents, spouses and friends of patients attending the event.

## Looking back and moving forward

**Rivka Plaut**

It was finally time to do what I had come here for—to thank all my nurses for doing such an amazing job. I knew that when I was a patient here, I was really sick and so I obviously didn't look very good and I felt like my nurses deserved to see the end result.

I wanted their vision of me to change from a girl who was extremely ill and dying to a girl who was healthy and very much alive.

As the elevator stopped at the 9th floor, I had so many different feelings and emotions going on at once. It was so surreal to be back there and to see everyone after so long. Thank G-d, so much had changed for me, but not much had changed on 9 south. I started walking down the hall, and right away, I remembered the first time that I ever walked down this same hallway. I didn't know what to expect, and I never imagined how this place would become my home away from home.

As I peered into one of the empty patient rooms, I remembered the countless days I had spent in there. I was shocked at how different two worlds can be. When I was a patient here, my entire world was the hospital. My outdoors were the garden, my "shopping" was the gift shop, etc. But coming back now, it is so crazy to see how small my world really was back then. I was so oblivious to what was going on outside of these four walls.

But after having gone exploring and touring all over Boston, the hospital seemed like such a small part of a bigger picture, but when I was here, it was my whole world.

I realized that this was also a comparison to my life. When I was stuck here in my own world, my entire life was all about being sick and it seemed like there was nothing else - that was all I could think about. Now, looking back, I can see that being sick was just a part of a much bigger picture- there is so much more! After being a patient there so much and becoming so sick, I began to feel safer on 9 south than in my own house. I knew that if anything happened, my doctors would try to treat me and my nurses would be there to hold my hand (literally) and take care of me, where as if I was at home, I would panic and freak out that the worst was going to happen.

Saying hello and thanking my nurses was the greatest part of my trip.



*Rivka Plaut and Kayla*

The way that they looked at me in amazement made me feel like all this had been worth it. I didn't realize how much they had all done for me until I was put in other hospitals all over the country.

I don't know the reason why I had to spend so much of my early twenties in a hospital, but I am happy that it was in Boston Children's Hospital. I am so lucky that they took care of me until it was time for me to get my new lungs. I said my good-byes to everyone and as I walked out of the revolving doors, I finally felt that I had closed the book on that part of my life, but only after I got to add the happy ending. Boston isn't only a place that is associated with sickness and dread anymore—it is now filled with good memories too. I hope that many more patients will be able to experience their own happy endings too.



*Leah Schwanke*

## Simplicity

**by Leah Schwanke**

The sun rises in the morning

The moon rises at night.

A baby cries when it is hungry

A moth is drawn to the light

How can some things be so

Simple while others cause a scare

How can people change

From night to day

Without a care

Why isn't it all that simple?

Even if it takes a while

To see what's in the mirror,

Crack a simple smile.

## Living with CF is not easy...

**Jeff Gaudet**

On the morning of April 17, 2000, my wife Lori and I celebrated our seventh wedding anniversary. Later that day our then three year old daughter Josilyn was diagnosed with cystic fibrosis.

"Finally," we said, "a diagnosis. Now how do we fix it?"

"Hold on," said the resident that had given us the sweat test results. "Let me get my boss." When we learned what we had and what it meant, you can imagine our dismay. Within an hour my wife and I would be learning chest physical therapy. I was taught one handed because two hands would not fit on my child's back. Some hours later, with a stack of prescriptions in hand and a dizzying array of instructions, numb and in disbelief we were ready to head home. "Aaah, when do we start PT?," I asked. "How long will it take you to get home?," was the reply, "about a half hour," I said. "That will be fine."...Whack, reality.

Since that day our life has not been the same. Each day there is chest physical therapy, antibiotics, pills and more pills, enzymes, nebulizers, inhalers, dietary supplements and overnight feeds. We were ultra compliant, we worked at it, Josilyn worked at it. However, the hospitalizations came anyway. From 2001 to 2004 we spent weeks upon weeks in the hospital. Dr Henry Dorkin and his team with a huge assist from my wife Lori, changed this

and adjusted that, increased one med and decreased another. A fundoplication and surgery for a permanent feeding tube (aka a Mickey Button). We adjust and communicate, adjust and communicate and test and observe and comply and then, do all that again. Viola, they figured it out. No hospitalizations from 2005 through most of 2011.

There have been stretches of good health and periods of adjustment. However, "team cheeka" our daughter's nickname and one we have, with respect, given to Dr. Dorkin's team and all those involved in Josilyn's care, stay vigilant and keep her healthy. Even with all that CF is a moving target. This school year had been rough for Josilyn, gastro intestinal issues had her sick and fatigued and out of school about 60% of the time since the year began. However, a quick "clean out" at Boston Children's followed by two more weeks of IV antibiotics at home and, she has not missed a day since Christmas. Adjust and adapt and always comply with treatment.

As for Josilyn, she is now 14 and is remarkable. To look at her she is the picture of health. She never, ever complains. She has missed a lot of school but manages to keep up with her classmates anyway. She is an honor roll student and an extremely active teenager. She has a spirit that warms and touches everyone she meets and most do not know she has CF. We have watched her blossom from a shy, malnourished, introvert a bit embarrassed about her condition, to a beautiful,

confident young woman. Josilyn thinks nothing about going to the mall, to a movie or out for a walk with her 16 year old sister, Jennifer, and her friends while carrying a bag containing a pump hooked up to intra venous anti biotic. She has taught them the S A S H protocol and together, without her parents being present, they handle IV med changes as needed. Her willingness to share some facts of her life with CF has fostered relationships with people who do not expect or allow her to take time off from her treatment regime.

On Tuesday, January 31, the FDA approved Kalydeco, the first and only drug on the planet that treats the root cause of CF. This is a game changer for her. It is not a cure but it should dramatically improve the quality of her life. We are excited and anxious. However, it is with some trepidation that I speak of our good fortune. We understand that for 96 percent of the CF community the day did not carry the same weight as it did for us. We know the news brings hope to them, we know they are happy for us. However, the percentages tell us most CF families will not have the same feelings we do. We understand and we extend our continuing prayer, hope, admiration and off course our fundraising efforts.

Living with CF is not easy. On the other hand, as a family we have a depth of love, an appreciation for life and for each other that we otherwise may not have known. God has blessed us.

*Liz Collette, Jennifer Gaudet, Josilyn Gaudett and Carlye Bertwell*



## Contact Information

To receive updates from the CF center via email go to [cfevents@childrens.harvard.edu](mailto:cfevents@childrens.harvard.edu) and add your name and address.

### For information on local CF events:

[cff.org/Chapters/mass-ri](http://cff.org/Chapters/mass-ri)

### Hospital Phone numbers:

Appointments: 617-355-1900 option #3

Nursing line: 617-355-7018

Prescription line: 617-355-7078

Pulmonary Function Test: 617-355-7510

Page Operator: 617-355-6369

Hospital Main Number: 617-355-6000

## Great Strides team

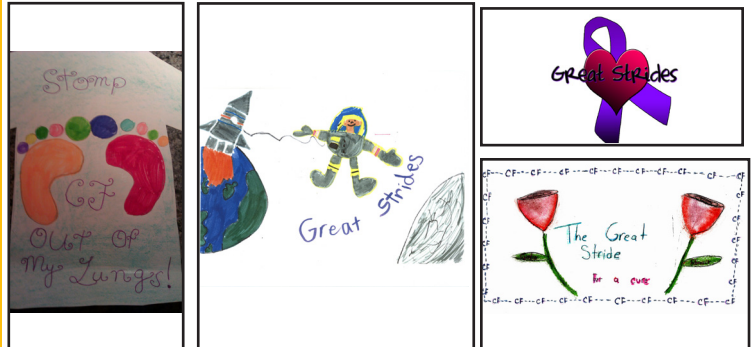
This year Ashish George and Katherine Koniaries are organizing the Boston Children's/Brigham Women's CF Center Great Strides team! We invite you to join the team and help to raise much needed funds to support cystic fibrosis research. You will also enjoy food, fun, music at the Great Strides event at the 2012 Boston - DCR Artesani Park site on 05/20/2012!

Thank you to all of the artists who submitted designs for our team tee shirt. The winner of the contest is: *Bianca Augeri*



### Runners up:

*Clockwise: Grace O'Leary, Ella Hannon, Zack Sullivan and Taylor Joron.*



News Wire

CYSTIC FIBROSIS