

CYSTIC FIBROSIS



News Wire

VOLUME TWO - NUMBER ONE - Winter 2010

We have just returned from the North American CF Conference (NACFC) in Minneapolis, and are pleased with the progress being made on various fronts. Almost 20 members of the CHB CF Center went to the meeting, with Center representation at numerous discussions and poster presentations. Anne Gould, Tom Martin, Erin Leone, and Greg Sawicki all were highlighted in presentations on airway clearance, airway infection, research coordination, and quality assurance. The CF Center was awarded the Foundation Quality Improvement Award for 2009, based upon the efforts led by Greg Sawicki and Ahmet Uluer.

Clinical trial interim results were presented for our edification on various topics, including early information concerning CFTR potentiation (Vertex trial) and best methods for early eradication of Pseudomonas (EPIC trial). Frank discussions on the best methods to choose antibiotics for intravenous and oral therapy led to a great deal of re-evaluation of how we choose antibiotics to treat our patients for exacerbations. Advances have led to our increased appreciation of how critical nutrition and vitamin D are for good lung development, growth and health.

It goes without saying that the collaboration with our patients and their families is a major reason for our success. Without the many volunteers to take part in clinic trials, many of the medications of the past (Pulmozyme and TOBI, to name two) would not be available for our patients today. Without such continued support and participation, the next generation of CF medications (which may dramatically alter the natural history and prognosis of Cystic Fibrosis) will not be developed. We have won many battles but have not yet won the war that all of us hope will conclude successfully and victoriously.

On behalf of all of us here at the CF Center, thank you for your support. We look forward to many more successes in 2010.

Hank Dorkin MD

Associate Chief, Division of Respiratory Diseases
and Co-Director, Cystic Fibrosis Center

The Cystic Fibrosis (CF) Center at Children's Hospital Boston was recently honored with the Cystic Fibrosis Foundation's Quality Care Award for 2009. The award recognizes outstanding quality in processes and accomplishments. The CF Center was recognized for sustaining quality improvement work that improved outcomes in patient care. The CF Center actively involves patients and families in identifying, designing and implementing improvement efforts.

Pictured with the award plaque are (left to right) Kate Barnico, RN, nurse coordinator, CF Center; Ahmet Uluer, DO, director, Adult Cystic Fibrosis Program; Gregory Sawicki, MD, MPH, associate director, CF Center; Hank Dorkin, MD, associate chief, Division of Respiratory Diseases, co-director, CF Center; Jacky Cortes, RN, Clinical Coordinator, Pulmonary Clinic; and Ann Motl-Taylor, clinical administrative coordinator, Pulmonary Clinic.



TAKING STEPS TO CURE CYSTIC FIBROSIS

GREAT STRIDES is the CF Foundation's largest fund-raising event. It provides opportunities for all people within a local community to get involved forming teams through their workplace, through clubs and

organizations or with friends and family. Walk day is a fun, family-oriented celebration that includes a healthy 10 kilometer (6.2 miles) walk, Kids' Corner activities, food, and festivities that participants look forward to year after year. Look for Children's Hospital/ Brigham and Women Hospital CF Center staff members as they walk to help find the cure.

Sign up today!
www.cff.org/Great_Strides

Team Leaders:

Amanda McGeachey and Ashley McCarthy

My Story

By Noam Liam



September of 2006 was when I hit the lowest PFT scores of my life. It was approximately 2 weeks after being hospitalized for the first time. Knowing I was one of the lucky ones who hadn't been hospitalized until after I turned 21 didn't help me from feeling down about this. Mentally I was not OK with how things were going; the medicine which doctors hoped would help seemed useless - and I decided it was time to do something dramatic in order to take control of my life. December of 2006 I started lifting weights every day for 1 hour, 5 days a week. No exceptions, no

excuses, this became priority number one. I needed to be in the best shape of my life. It was the first time I had a feeling of pure dedication.

I cannot stress enough the difference that eating well and exercising has done. Not only do I look better physically, but I have more energy and I have gained weight. My PFT scores went up dramatically, 20%. This was with the addition of the salt-saline inhalation into my health routine, but there was no doubt that my physical shape helped as well. The stronger my chest became, the easier it was for my lungs to do the work they needed to do. I know these sounds like another skeptical story but these are in fact true results.

Exercise:

I didn't want to spend countless hours at the gym. I used a free online training series. It is a daily/weekly/monthly exercise regiment. At first I followed it by the letter. After several months I added my own variations to accommodate my schedule better. Trust me it worked for me only 1 hour a day and it has been very effective.

Diet and Supplements:

I wanted to make sure every second and

every ounce of energy I was putting into this would not go to waste. I also was not about to pump a lot of chemicals and weird substances into my body so I did a lot of reading on the supplements that were needed when lifting. So I researched and found diet supplements that would help me. As far as supplements go, I take a protein powder supplement

SCANDISHAKE Probably the most important here, this helped me pack on the pounds!

1 Packet scandi-shake; a little bit of decaf instant coffee and whole milk. This was a shake I would have every single morning (Mon-Fri) and added 750 calories to my diet. I have a protein bar that adds another 150-200 calories and I eat 3 meals a day.

It has been a while since Dec '06, and I stopped lifting at times, but my PFTs reflected it. It is very important to keep at it and stay in shape. I'm normally not very outspoken about CF - but I do honestly believe the combination of exercise, salt-saline, eating healthy and a dedicated attitude is what has helped me stay in such great health.

RESEARCH

Why is this research being done?

People with cystic fibrosis (CF) have to do many treatments each day.

We know that patients sometimes miss their medicines or treatments because they forget or are too busy.

We are learning which tools and strategies help CF care teams communicate better with their patients and improve their patients' CF management.

What will happen if you join the iCARE study?

- You will complete an assessment that will take about 45 minutes. The assessment includes a survey and an interview.
- You will repeat the assessment at 12 and 24 months after joining the study.
- You may be asked to complete problem-solving sessions with a CF team member as part of your clinical care.
- You will be compensated for your time.
- There are no investigational drugs or devices in this study.



Are you eligible for this study?

You (your teen) may join the iCARE study if you (your teen):

- are diagnosed with CF
- are 11 to 20 years old
- attend an accredited CF care center for regularly scheduled clinic visits
- have been prescribed at least one of the following medicines for the past 6 months:
 - Azithromycin
 - Hypertonic saline
 - Pulmozyme®
 - TOBI®
- Inhaled compounded tobramycin
- have consented to provide data to the CFF Registry.

If you are interested or want to learn more, please feel free to get in contact with the research assistant, Nathan Demars, at: 617-355-7248 or email him at: nathan.demars@childrens.harvard.edu

COLLEGE PLANNING

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

Planning for college can be a challenging and exciting time. Identifying and visiting potential schools, completing applications, exploring possible health related accommodations (for you or for your student), and seeking out financial aide resources are some of the tasks you are facing. Every student has his/her own special circumstances which may impact upon his/her choice of a particular college. Students living with CF may in addition be asking: what factors, such as location/terrain/health services, do I need

to consider when I look at a particular college, how do I decide what type of housing is best for me, how am I going to fit in all of my treatments while still "having a life" at school, what happens if I'm sick and have to miss class? Your CF SW can help you figure out how you might handle each of these situations and can in addition provide you with resources on potential scholarships and on how to access the college/university office for students with disabilities or health needs. She also will be sending you a college

planning packet.

Deciding on a "good fit" school and figuring out how you will incorporate your CF care and staying healthy into your day are two primary tasks/goals. Being prepared by utilizing all of the resources available to you as you get ready for school will ensure an easy and smooth transition and help you make the most of your college experience.

patient family ADVISORY council

As we close out 2009 we can reflect on our accomplishments and look to our future with renewed focus and commitment. This year we delivered a quarterly Newswire, provided a "Med School 101" event and hosted an evening at the Marriott to review the North American CF Conference updates.

The next lecture topic: March 6th "Keeping Your Lungs Healthy". Come and learn about; PFT's, (what do the numbers mean); chest x-rays; airway clearance and home equipment.

The PFAC continues to meet monthly and welcome new members; please contact us. We meet the third Tuesday of each month at the Children's Hospital Waltham facility @ 6:00 pm. Have an idea? Please email us or speak with your social worker to make a connection and be a part of PFAC. Email Amory Julian at amoryjulian@comcast.net or Ellen Brown at ellbrown@yahoo.com.



healthy habits

Cleaning the Nebulizer Parts

- With a new paper towel, wash the inside and outside of the nebulizer parts with water.
- Be careful not to damage any of the parts.
- Throw the paper towel away, and then rinse the nebulizer parts with water.
- Clean the nebulizer right after it is used to keep the medicine and debris from drying. Once debris dries, it is difficult to wash off.
- You can clean the nebulizer parts in an automatic dishwasher, if the nebulizer's manufacturer's instructions allow.

Disinfect the Nebulizer Parts Daily

Do not use VINEGAR. Vinegar is not strong enough to kill the germs a person with CF might get. Instead, if the manufacturers' instructions allow, disinfect the nebulizer parts using one of these options:

- Boiling for 5 minutes
- Microwaving (in water) for 5 minutes.
- Dishwasher, if the water is hotter than 158F for 30 minutes.
- Soaking in a solution of 1 part household bleach and 50 parts water for 3 minutes.
- Soaking in 70% isopropyl alcohol for 5 minutes.
- Soaking in 3% hydrogen peroxide for 30 minutes.
- Do not use a nebulizer that cannot be disinfected using one of these options.

Rinse the Nebulizer Parts

- If you rinse using bleach, isopropyl alcohol or hydrogen peroxide, rinse all parts well.
- Use sterile water for the final rinse.
- **DO NOT USE WATER FROM THE FAUCET, BOTTLED, OR DISTILLED WATER**
- You can make sterile water by boiling it for 5 minutes.
- Use this water once then throw out.

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- If you disinfect by boiling, you do not need to rinse the nebulizer.

Air-dry the nebulizer Parts

- After the final rinse, drain the parts on a clean surface covered with new paper towel.
- Replace wet paper towels with dry ones and fully air-dry all parts.
- Remember, germs will grow on anything that stays wet.
- Follow the manufacturer's care and cleaning instructions for all equipment used for inhaled medicines or airway clearance.
- Some respiratory equipment may need to be cleaned but not disinfected.

This information is based on "Infection Control recommendations for Patients with Cystic Fibrosis: Microbiology, Important Pathogens, and Infection Control Practices to Prevent Patient-to-Patient Transmission" by Lisa Saiman MD, MPH; Jane Siegel, MD; and the participants of the CF Foundation's Consensus Conference on Infection Control



AIRWAY CLEARANCE

The CF Foundation recommends that airway clearance techniques be reviewed at least once on an annual basis. Anne Gould can meet with patients and families in clinic to review the various methods of airway clearance, home programs for airway clearance, guidelines for regular exercise programs, and to answer any questions you have about physical therapy. To schedule time to meet with Anne please call (617)355-6079.

FLU UPDATE

The flu spreads easily through person-to-person physical contact and through the coughing and sneezing of infected people. There are a number of everyday actions that you can take to stay healthy:

- Cover your nose and mouth with a tissue when you cough or sneeze. Throw the tissue in the trash after you use it.
- Wash your hands often with soap and water, especially after you cough or sneeze. Alcohol-based hands cleaners are also effective.
- Avoid touching your eyes, nose or mouth.

- Stay home if you get sick. Limit contact with others to keep from infecting them.

H1N1 Flu vaccine: According to the CDC guidelines, people with CF are considered a high-risk group for developing severe influenza disease and should receive the H1N1 influenza vaccine because of chronic pulmonary disease. The CF Foundation's Influenza Advisory Group recommends that close contacts of people with CF (including household members without CF) should also receive the H1N1 Flu vaccine. As a reminder children under the age of 10 years of age should receive a second H1N1 vaccine at least 28 days after the initial dose. The CDC Web site: <http://www.cdc.gov/h1n1flu/vaccination/dosespaceing.html>

living with cystic fibrosis



by Amanda Healey

Living with Cystic Fibrosis comes with both highs and lows. Well, mostly lows. But in those difficult times, I try to focus on the highs in my life. I can't be upset all of the time; I'm the kind of person that likes to be happy and is always smiling. I'm the kind of person that believes laughter really is the best medicine. However, there is still no way around the treatments, medications, and doctor appointments that come with CF. When I was little, the worst thing was going into clinic. I've always hated doctors and needles. Just having my blood pressure taken made me anxious. In the past few years, things have changed for the better. I know how to calm down when I get anxious, and I know how to hold in my tears until after the doctor leaves the room. When I go into clinic now, there is a reason why I'm not afraid anymore. Brandon, the phlebotomist who works in the pulmonary department, has made a big impact on my attitude towards clinic. When I walk up to the front desk, I am greeted with a smile from Brandon as he quickly looks for a clean room where all the clinic regulars will check in on me. The first task for Brandon is to check my vitals, and he never seems to run out of things to say: "How are you doing? How's your brother? Is he being nice? Tell him I say hi!" And he always knows how to make me

laugh. For instance, I once brought in a magazine to read while I waited – you know the gossipy kind of magazine that is mindless and helps time pass. Brandon came in and swiped it out of my hands and started flipping through it, commenting on the pictures of celebrities. He made some really silly comments, confusing Ashley Tisdale with Brooke Hogan! Then he tossed it back to me and walked out of the room. My mom and I just looked at each other and started to laugh. Another time, he had to draw blood from my brother, who also has CF. He didn't have any of the little needles, so he tried to make light of the situation by saying, "The bad news is that I have no little needles. Do you want to hear the good news? I just saved a bunch of money by switching my car insurance to Geico." Personally, I found that funny. I also learned from him that if I have a distraction while getting a shot or blood drawn, then it doesn't hurt as much. One day my doctor was taking a long time to see us, so Brandon came in and put on a movie for me. I hardly noticed when he came in and took some blood. I was just focused on the movie (which was Madagascar - yes, I was so immersed in the animated Madagascar movie, that I did not notice a needle in my arm!) From then on, I have taken my iPod and headphones into every doctor appointment to help me relax when I get anxious, and to distract me when needles are near. Brandon is always there to make me laugh even when I feel like crying out of frustration. Now, instead of absolutely dreading and having anxiety about going to clinic, I look forward to the ways Brandon will put a smile on my face. He cares about all of his patients and I don't think I could thank him enough for how he has helped me live with Cystic Fibrosis.

SPOTLIGHT ON TRANSITION

Greg Sawicki, MD, MPH

There is a new and exciting challenge facing the cystic fibrosis (CF) medical community today: how do we successfully treat the growing number of adult patients with CF? In 2007, the Cystic Fibrosis Foundation reported that the number of adult CF patients was growing at such a rate that it would soon exceed the number of pediatric CF patients in the US. This is an extremely promising finding, but it also represents new challenges for CF patients, their family members, and CF care teams.

Why transition?

At first, the idea of transitioning from a pediatric CF center might not make much sense. After all, the quality of care can be top notch and trusting relationships are created with care team staff. However, adult CF patients face an increasingly different set of challenges than children, including:

- New health challenges: Adults patients are often faced with new CF and non CF-related health issues that can include diabetes, serious infections caused by drug-resistant bacteria, reproductive issues, etc.
- Evolving personal challenges: The pressures of daily life as an adult (college, family planning, career development, marriage, raising children, etc.) may require a unique approach in terms of support and guidance.

Today, dozens of adult CF care centers have been set up across the world to serve the specialized needs of the adult CF patient.

When should transition planning begin?

Transition to an adult CF care center is an important milestone for any CF patient and family. It does not simply include moving on to a new hospital, clinic, or doctor. It is a process that takes several years and involves a partnership between patients, families, and clinicians. While there is no “perfect” time to transition to adult care (it varies based on the individual), developing skills necessary to facilitate transition should begin early in adolescence.

An Overview of Transition Skills for Children and Teens

Age	Transition Skill
Children (6-11 years)	<ul style="list-style-type: none">• Count out enzymes daily• Learn the names of each medication• Maintain a symptom diary
Pre and Early Teens (12-15 years)	<ul style="list-style-type: none">• Learn what each medication treats and why it was prescribed• Keep an appointment calendar (electronically or in a pocket organizer)• Ask questions of the doctor and care team at each clinic visit• Fill out medical history forms• Take ownership for cleaning medical equipment and supplies
Teens (16-18 years)	<ul style="list-style-type: none">• Understand current insurance coverage• Organize transportation to and from doctor visits• Communicate with the CF care team independently at scheduled visits• Keep track of current treatment regimen and take medicines as prescribed by the CF care team• Take responsibility for ordering prescription refills

How do I begin the transition process?

A great first step is to begin transferring age-appropriate treatment responsibilities to a young patient. Over time, it is important to provide increasing levels of responsibility to the child and teenager. At our clinic here in Boston, we are in the process of developing a simple checklist tool that helps CF patients (as well as our team) understand the critical aspects of the transition process and highlight areas of need. The chart below provides some examples of age appropriate transition skills.

Are there any transition tips or “best practices” I should be aware of?

It is important to remember that transition is a process. Like any significant life change, it can be exciting and invigorating, and stressful and scary. Chances are, you will experience one or all of these emotions over time. Here are a few tips to help you as you embark on transition to an adult CF care center.

Tips for a Successful CF Transition:

1. Stick with it. Transition is a long-term process. Acquiring the transition skills necessary for success will not happen overnight.
2. Communicate. Clear, open, and honest communication between the patient, family, and care team is essential.
3. Compile a medical summary. Develop a complete medical summary from your pediatric team that can be shared with the adult care team.
4. Collaborate. Joint visits with your pediatric and adult care teams can ensure continuity in medical care.
5. Transition is a family affair. Patients and parents must work together and be sensitive to each other’s feelings during this time.
6. Remember, you are not alone. Transition is a critical phase in the life of any CF patient. Many CF patients have successfully navigated the transition process, and like you, hundreds more are working toward a successful transition today.

NUTRITION UPDATES

Kristen Leavitt, RD, IDN | Erin Redding, RD, IDN |



Nutrition plays an important role in the treatment of CF. People with CF need more calories, and this is often complicated by decreased appetite and their body's difficulty absorbing all of the nutrients they eat. Although it may be difficult for patients with CF, maintaining a healthy weight is associated with improved lung function and longevity. The CF foundation recommends patients maintain a body mass index (BMI) or weight for length at or above average to help the lungs work the best that they can. For patients less than two years old the goal is a weight for length at or above the 50th%ile for age on the growth chart.

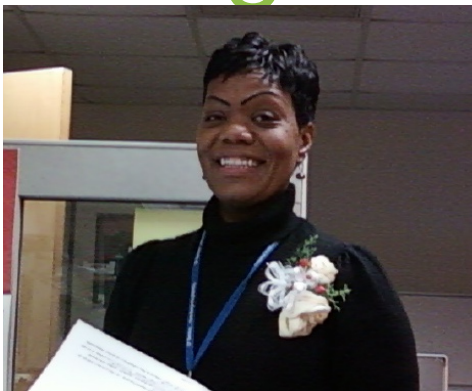
For patients two years and older, the goal is a body mass index (BMI) at or above the 50th%ile for age on the growth chart.

Registered dietitians are members of the CF care team who are most responsible for monitoring growth and nutritional status and providing up to date nutrition recommendations, ongoing counseling, education, and support around feeding and nutrition issues. Frequent monitoring and early intervention by the dietitian have proven to help with appropriate growth and weight gain in CF patients, in turn improving their lung function and overall health. The CF Foundation recommends that all patients be assessed by a registered dietitian annually. Patients who fall below the 50th%ile weight for length or BMI for age may benefit from seeing a registered dietitian more frequently.

We would like to take this opportunity to introduce ourselves as the new registered dietitians here at Children's Hospital Boston CF Center. We are passionate about nutrition and CF and understand that nutrition is an important part of your overall care. Eating the right diet can often be challenging, and many patients and families find it difficult to balance the demands of nutrition with other aspects of their treatment. Our role is to work with you to make nutrition goals achievable and tailored to your individual needs and lifestyle. We acknowledge that there is more to CF nutrition than simply eating a high calorie diet and hope to serve as your nutrition expert in all areas of CF nutrition care. We look forward to meeting and working with you in the future. For more information please contact:

Kristen.leavitt@childrens.harvard.edu or
erin.redding@childrens.harvard.edu

congratulations



Congratulations to Renetta Johnson, Administrative Associate IV. Renetta has been awarded the Children's Hospital Black Achiever Award for 2010. Other recent award winners include Employee of the Month winners Arelis Munoz, surgical scheduler (August) and Larry Rhein MD (November).

CONTACT INFORMATION

Phone numbers:

Appointments: 617-355-1900 option #3

Nursing line: 617-355-7018

Prescription line: 617-355-7078

Home IV Program: 617-355-6499

Pulmonary Function Test: 617-355-7510

Page Operator: 617-355-6369

Hospital Main Number: 617-355-6000

UPCOMING EVENTS CF EVENTS

January 30th

CF Dinner Hyatt Harborside

March 6th

"Keeping your Lungs Healthy"

Dr. Hank Dorkin and Anne Gould PT

May 15th/16th

Great Strides CF Walk

News Wire

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