Hello and Happy New Year!

Welcome to another edition of the Children’s Hospital Boston CF Center “CF Newswire.” Hope everyone has finally shoveled out and is ready for spring!

It was great to see so many of our families at the CF Center’s annual dinner in November. It was a very successful evening with 150 family members and staff in attendance. Families had time to socialize with other families and staff as well as learn about ongoing initiatives within the CF center. The CF Center leadership discussed latest developments in CF as well as updates from both the pediatric and adult programs.

This past October, 24 members of the CF center team attended the North American Cystic Fibrosis Conference in Baltimore. Several members of our team presented in symposiums at the meeting. Greg Sawicki MD spoke on strategies to approach MRSA; Alicia Casey MD presented at the pediatric fellows’ case conference; Ahmet Uluer DO presented data on “Urinary Biomarkers for Early Detection of Nephrotoxicity in Cystic Fibrosis”; and Keri Sullivan NP presented data on our ongoing home IV therapy program. In addition, members of our multidisciplinary teams were able to join discussions in roundtable sessions, workshops, and other collaborative meetings during the conference.

Over the past several months, we have welcomed several new staff members. Ashish George and Beatrice Duvert have joined the clinical research team. The new PFT technicians are Daniel Goulette, Trevor Wright, and Walker Del Aguila. Courtney Madden RN joined the clinic nursing team and Leah Frain FNP joined the adult CF team. Robin Welcher RD, LRD is the primary inpatient nutritionist at Brigham and Women’s Hospital. We have also established formal collaborations with 2 physicians at Children’s Hospital: Elizabeth Yen MD is a pediatric gastroenterologist with a special interest in GI manifestations of CF. She will be available to see patients on Farley 4 for consults. Georgina Garcia MD is a child psychiatrist who will be the primary consult attending for our pediatric inpatient service.

The Patient Family Advisory Committee has been busy planning upcoming events. The group meets monthly at the Children’s Waltham Hospital site on the third Tuesday of the month. We are a group of patients, families and staff working together to improve care at the CF center.

Do you have a suggestion, story, photo or an accomplishment you would like to share with the “CF Newswire”? Email to cfevents@childrens.harvard.edu.

Kate Barnico, RN, BSN
Cystic Fibrosis Center Coordinator
Children’s Hospital Boston

NINA’S CHANCE MEETING

By Doreen M. Cummings

On a warm summer’s evening this past August, our family had the best experience at the Beach Plum Inn on Martha’s Vineyard. Our family travels to the Vineyard each summer, during the last week of August to enjoy swimming, fish filled meals, long days at the beach and sunshine! This year, the Obamas happened to be visiting the Island at the same time, and we all happened to pick the same place to have dinner on Friday Night!

(Continued on final page.)

AIRWAY CLEARANCE TECHNIQUES

Airway clearance techniques (ACTs) are treatments that help people with cystic fibrosis (CF) stay healthy and breathe easier. Clearing the airways reduces lung infections and improves lung function. There are several different techniques that can be used to help clear secretions; such as; chest physical therapy, the Vest, Flutter or Acapella, and/or exercise.

ACTs are often used in conjunction with other CF treatments. Your CF care team will help you choose the best ACT and therapies. It is recommended that patients formally review ACT each year with a physical therapist. We will try to schedule this at the time of one of your clinic appointments. Anne Gould PT will review and evaluate your current ACT and exercise routines. Appointments can be made by calling 617-355-6079.
Walpole High girls’ track coach Conan Cashman and good friend Eileen “Meg” Henneberry of Westwood give the thumbs up to a road race three years ago in her fight with Cystic Fibrosis.

An intercollegiate athlete, she played lacrosse at Northeastern University and received an engineering degree graduating magna cum laude. Henneberry has a strong competitive drive and work ethic, according to those who know her.

“On my two year ‘Lungversary’ I said enough! Start running! You have been sitting on the sidelines way too long and being lazy,” said Henneberry. “Use those lungs. Use this gift of life from your donor and appreciate it.”

Walpole resident Dawn Freiberger, a nurse and the lung transplant coordinator at Children’s Hospital in Boston said, “It is very hard for her to not just finish the race but do it in a good. I wasn’t fighting anymore, I was breathing like a normal person,” she said.

Since she had been in a coma, her entire body atrophied. The muscular tone that she had been so proud of as a college athlete was gone. “I couldn’t sit up, certainly couldn’t stand. I never took a deep breath outside, ” said Henneberry.

“The hearing situation was easier to deal with than I thought, so what if I can’t hear. I can’t breathe? I can’t walk?” said Henneberry.

“My health is the best it has ever been,” said Henneberry.

Henneberry said that even after a lung transplant operation, it is “like trading one disease for another” due to frequent, ongoing medical complications. She said that the new lungs will not be affected by the disease. She said Henneberry has had a “rocky road” after the lung transplants but that she is doing well now. “She is one of the most motivated patients I have seen. She takes great care in keeping herself healthy.”

After the transplant operation, Henneberry said she was just 95 pounds and was so wasted she could barely eat or even change the TV channel at the hospital. “I would have dreams of running everywhere, playing lacrosse, sprinting in to make a goal and I would wake up, oxygen mask on, almost unable to get out of bed on my own accord. It was depressing,” said Henneberry.

“A small gift from one person can change a whole world for another person,” said cashman.

Henneberry said she was just 95 pounds and was so wasted she could barely eat or even change the TV channel at the hospital. “I would have dreams of running everywhere, playing lacrosse, sprinting in to make a goal and I would wake up, oxygen mask on, almost unable to get out of bed on my own accord. It was depressing,” said Henneberry.

“A small gift from one person can change a whole world for another person,” said cashman.

Henneberry said she was just 95 pounds and was so wasted she could barely eat or even change the TV channel at the hospital. “I would have dreams of running everywhere, playing lacrosse, sprinting in to make a goal and I would wake up, oxygen mask on, almost unable to get out of bed on my own accord. It was depressing,” said Henneberry.

“A small gift from one person can change a whole world for another person,” said cashman.

Henneberry said she was just 95 pounds and was so wasted she could barely eat or even change the TV channel at the hospital. “I would have dreams of running everywhere, playing lacrosse, sprinting in to make a goal and I would wake up, oxygen mask on, almost unable to get out of bed on my own accord. It was depressing,” said Henneberry.

“A small gift from one person can change a whole world for another person,” said cashman.
Living with CF is about living with hope and living about some denial, about living with optimism and about living with possibility. Everyone with CF and every family member touched by CF knows that life is more tricky and at times more challenging because of cystic fibrosis. Denial as a mechanism for coping can be useful and healthy or fruitless and harmful. Healthy denial allows you to get up and keep living, to face the realities of the day, to do what you have to do, to enjoy the moment, and to plan for the future. Unhealthy denial can cause you not to set daily CF care as a priority, nor make or keep CF clinic appointments, nor continue treatments, nor be vigilant about diet and exercise, and delay in getting medical attention. While the intensity of denial may be altered when you or your family member is confronted with a change in health, attitude and perspective can help you get through these times.

Hope is likewise a critical mechanism for coping. CF is a diagnosis and not an illness or sickness. When problematic symptoms arise, your CF care team partners with you to treat these episodes. Information on CF is ever-evolving and Internet written materials may often be out-of-date. You should always feel comfortable asking questions of your health care team about new advances in research and treatment of CF and about resources for you and your family members.

Everyone with CF are living long and productive lives. Daily treatments do make a difference, enhancing and improving quality of life and increasing energy and strength so that you (or your family member) can accomplish the things you want and need to do. At times, however, even when being conscientious about your daily care, a CF exacerbation may occur and it may be no one’s fault. Putting hope into your life is working hard to make a difference, enhancing and improving quality of life and increasing the most calories: protein, carbohydrate or fat. Do you know the answer? It is fat! Fat has 9 calories for every gram eaten where protein and carbohydrate have only 4 calories per gram. Did you get the answer wrong? Don’t worry. Most people asked did! When parents were asked this question they got it wrong 87% of the time. Our adolescents got this question wrong 89% of the time. Because of this, we thought we would dedicate this Nutrition article to the basics of CF nutrition. As many of you know, it can be very challenging to eat large amounts of food at a sitting. To help those with CF consume the calories necessary for either weight gain or maintenance, meals and snacks must be packed with calories. As stated above, fat contains more calories than protein or carbohydrate, and can often make foods taste better. Foods high in fat, such as cheese, milk, ice cream and meat, are also good sources of protein. Protein is important for building muscle and helping the body prevent and recover from illness or infection. Foods high in fat can also be high in carbohydrate (think baked goods and granolas), which provide the body with energy and fiber.

WAYS TO BOOST CALORIES

Although fruits and vegetables are important to include in the diet, it may be necessary to limit the amounts of these foods, as they will fill up the stomach without providing many calories. Adding sources of fat to these foods will increase calories and can make eating them daily a healthy possibility: Add butter, margarine or oil to breads, cereals, rice, noodles, potatoes and vegetables. Add sour cream to meat, potatoes, vegetables, and casseroles. Dress up fruit with cream cheese, whipped cream and chocolate sauce. Add extra salad dressing, nuts, dried fruit and cheese to salads. Added fat can also be used for meats and starchy to boost calories. Meats can be marinated with oils before cooking. Use creamy and cheesy sauces or gravies with meat, and casseroles. Mayonnaise can be added to sandwiches and the fillings used in them.

You can add fat to additional foods by using whipped cream on hot chocolate, fruit, pudding, pie and other desserts. Add syrup, jam, jelly or hard toppings to ice cream. Use peanut or other nut butters, or extra jam, jelly and honey on toast, bread, muffins, biscuits and crackers.

Full-fat dairy is a good source of fat, too. Prepare soup, cereal, hot chocolate and pudding with half-and-half or heavy cream.

Serve whole milk and other whole-fat dairy products such as cheese and yogurt. Add cheese to scrambled eggs, sauces, vegetables, soups, casseroles and salads. Lastly, try adding extra eggs or just egg yolks to sauces, casseroles and salads.

USING SNACKS WISELY

Children may complain during a meal that they are too full to continue eating. Rather than pushing your child to eat large quantities during meals, it is helpful to give high-calorie snacks in between regular meals. You may want to give snacks (“small meals”) more often throughout the day if your child often feels full and cannot eat very much during meals. Children that snacks are given at a certain time in between each meal rather than allowing your child to snack or “graze” throughout the day. This will cause preemptive fullness and lessens appetite prior to mealtimes. This concept applies to those with CF of all ages.

TIPS FOR HIGH CALORIE SNACK

Here are a few ways to make eating high fat foods easier:

• Make a list of high-calorie bedtime snacks for you or your child to choose from.
• Think of ways to turn lower-calorie snacks (such as crackers, apples and cereal) into higher calorie ones by adding peanut butter, butter or cheese.
• Keep nut butters, cheese and full-fat dairy stocked in the house for easy access.
• Find or create recipes for milkshakes and smoothies made with a combination of ice cream, powdered milk, cream, instant breakfast powders, yogurt, and fruit.
• Keep meal bars, granola, nuts and potato chips in the cabinet for high-calorie eating on the run.

This article provides a short list of ideas on increasing your caloric intake. We encourage you to make an appointment with one of the CF dietitians—either Kristen or Ashley—to discuss new, creative strategies for high calorie eating! We look forward to seeing you in clinic.

Have a CF nutrition tip or recipe? Share it with other patients and families at cfevents@childrens.harvard.edu.
I spent the last semester interning at Hospice & Palliative Care of Cape Cod, learning about the resources available for the many patients and families coping with serious illness. I saw that everyone has a story to tell. At every hospice event I went to, someone came up to me and shared their experience dealing with loss. I saw how their perspective on life and death gave them a special appreciation that echoed some of mine.

I believe that the most important things in life are the relationships we nurture. Death is uncontrollable; it can touch anyone at any time, without notice. I know that. There is not one day that I am not appreciating being alive.

I urge you to go out of your element and experience something or someone that makes you think differently. Life was not designed to be easy, but it is worthwhile. I live for experiences, for picnics with good food and wine, and moments when I don’t want to go home.

People live their whole lives working; only at the end might they consider what gave their life meaning. Every day you live, ask yourself: What are your goals for your life? What is important to you? I have been blessed because I know the importance of figuring out life’s meaning and then living accordingly.

I am not so different from you; I don’t look like a sick person. I could be sitting next to you at your office, or waiting on your family at dinner. Everyone is similar, with goals, fears, hopes and a lot of stress. We are all special. Including you! Life is a terminal illness for all of us. It might last 10 more years, or 30; you never know. I have had several friends die before I reached 30 and have same burdens. Every day as a time for happiness — regret just wasn’t a word in my vocabulary. They lived too briefly, but so unconditionally and with such passion that thinking of them makes me cry. I have had the pleasure of knowing amazing people who miss being close to others who have the same burdens. Every time I do something special, I think of them, and carry them with me as I continue on my life’s journey.

This month I am off to Italy to study photojournalism with Paul Daniele. I am looking forward to seeing the Italian culture and will return home to Cape Cod, to fulfill my dreams of a home and family, much like many others graduating soon. May we all find the hope and joys in each of our unique lives, for whatever time we each may have.

For many young people, May is a time for graduation and planning for the future. They look forward to getting out into the real world, their first job, meeting their life mates. These days especially, they are likely to worry about money, career, and expectations for success.

As I approach my own graduation from the Suffolk University program at Cape Cod Community College, I have a different perspective.

I was born with cystic fibrosis. I was baptized right away, not weighed and measured, because the doctors weren’t sure I would survive. As I was rushed down the hospital corridor to intensive care, my parents were very frightened. I grew up with constant medical treatment and the reality of a limited life expectancy always hanging over my head.

As you might imagine, this has been a huge burden for my family and me. But you might be surprised to hear that I also consider it a blessing.

Every birthday is truly a time for celebration. Most people worry about aging, but I yearn to experience graying hair and laugh lines circling my mouth. I don’t expect them. I can’t.

I am not so different from you; I don’t look like a sick person. I could be sitting next to you at your office, or waiting on your family at dinner. Everyone is similar, with goals, fears, hopes and a lot of stress. We are all special. Including you! Life is a terminal illness for all of us. It might last 10 more years, or 30; you never know. I have had several friends die from cystic fibrosis in the past few years and every time I wonder: why not me? Why am I stuck in this paradox of being sick, yet outliving others? Why do I feel like I’m the only one of us left? No one who is loved is ever truly dies. So long as their memories are alive, stories can be told and passed on, keeping their spirit with us.

Medication Tips

medications

- albuterol/Xopenex
- hypertonic saline
- Pulmozyme
- Airway clearance
- Inhaled antibiotics
- Inhaled steroids
One minute we were ordering our dinner and the next, there was a fever in the air as our President, and Mrs. Obama made their way through the restaurant and outside to view the gorgeous sunset. The Beach Plum Inn is one of the favorite spots in Chilmark for viewing the beautiful summer sunsets. Nina and her Aunt Buket walked through the small crowd and greeted Mrs. Obama who was full of warm energy and who posed for a quick photo. Then, Nina and Aunt Buket waited for President Obama to finish talking with someone to have their turn for quick chat. President Obama sweetly talked with Nina, and asked Nina if she enjoyed her dinner if she was swimming a lot during her vacation. Then, we all returned to our tables and had a wonderful meal.