



Boston Children's Hospital

Until every child is well™

Cystic Fibrosis Center



BRIGHAM AND WOMEN'S HOSPITAL



Cystic Fibrosis Newswire

Fall 2014

Fall is a great time to be outdoors in New England

AHMET ULUER, DO

Welcome to another edition of the Boston Children's Hospital and Brigham and Women's Hospital (BWH) CF Center Newswire.

We hope you had a great summer! Our cystic fibrosis team kicked off the summer at Great Strides in May. We had a wonderful turnout at sites all over New England. It was a great success! As a staunch proponent of exercise, this event is near and dear to my heart. It gets all of us outside walking and running, and exemplifies the importance of exercise in improving quality of life and respiratory health. It doesn't matter how young or old you are or how well or sick you might feel, exercise in some form should be a part of everyone's life. Our oldest patients with CF universally credit exercise and their increased activity level as a contributor to their longevity. We continue to offer exercise as an assessment and treatment at Boston Children's and Brigham and Women's. We would like to make exercise and fitness a permanent part of your life, so please ask us about how we might help make this happen at your next visit.

As our patient numbers grew, particularly among those over age 21, we needed to find room to expand. After turning down an offer to move our adults with CF to a nearby community hospital, we decided to split into two groups: those over 35, and those under 35. Our older group of patients will continue to receive their inpatient care at Brigham and Women's, while those younger than 35 have already joined the Weitzman Family Bridges Adult Transition Program, at Boston Children's. This program is designed to support the medical and surgical needs of young adults with congenital or acquired chronic childhood diseases, with a focus on transitional care. Our young adult CF patients have found a wonderful new home in the Weitzman Family Bridges Program and it will continue to improve over the next year or two before taking its final form. We will provide age-appropriate care along with transition readiness assessments to help promote independence. Exercise is heavily encouraged with options to use a treadmill or stationary bike. We also have small weights and yoga mats for you to use in between respiratory therapies.

Along with the changes to the adult CF program, we have also implemented an updated infection control policy, based on new Cystic Fibrosis Foundation (CFF) guidelines which are due to be published very soon. Many of the new recommendations are ones we implemented long ago. Some of the big changes at Boston Children's include wearing a mask when in public areas on campus (not while in a clinic room or hospital room), maintaining a distance of 6 feet between patients (previously 3 feet) and new restrictions on leaving your room at Brigham and Women's. Please ask us for details next time you come to clinic or are admitted to the hospital.

Our adult CF program is a close-knit community, and we are proud of our patients and families who work so hard every day to remain healthy. We will continue to stay ahead of the curve, providing you the latest treatment options, new clinical research opportunities, improvement in outcomes through quality improvement initiatives and more. Our multidisciplinary team of caregivers is here to support you.

REACT (Re-Education of Airway Clearance Techniques) is an educational program which we began in our adult clinic last year.

ANNE GOULD, PT

A consistent daily regimen of airway clearance techniques and treatments is critical to maintaining good long-term health for people with cystic fibrosis. Airway clearance is a combination of inhaled medications, techniques and devices. It may include bronchodilators, hypertonic saline nebs, Pulmozyme, chest physical therapy (percussion and vibration with postural drainage positioning, use of a vest, a PEP device, acapella, Flutter or Aerobika. We have found that many of our adult patients were taught (or their parents were taught) airway clearance techniques many years ago. This program is designed to re-educate patients in the importance of daily airway clearance treatments and review correct techniques. It is also an opportunity to discuss the multiple options for airway clearance and assist patients in overcoming barriers to performing daily treatments.

As an incentive to participation in this program, the first 50 patients are being given free parking. The program involves bringing your airway clearance devices to clinic and meeting with a physical therapist for about 45 minutes. If you are interested in participating in this program, please contact Simona Rits at 617-355-1923 to schedule an appointment.

Updates in the Infection Prevention and Control Policy

The Cystic Fibrosis Foundation has recently brought together experts in infection control and CF lung disease to update guidelines regarding infection control practices for all individuals with CF.

The guidelines include information on ways to prevent the spread of infections in clinics, in hospitals, at home and school and at CF Foundation-sponsored events.

At the CF Center at Boston Children's Hospital and Brigham and Women's Hospital, we have long-established infection control guidelines, which have included universal policies for all individuals seen in either inpatient or outpatient settings in our hospitals. We follow a practice of enhanced contact precautions regardless of infection status. These precautions include proper hand hygiene, use of gloves and gown by all staff having contact with a CF patient, having patients go directly to an exam room and not wait in a common area, and the use of the "blue pass" when sending a patient for testing (for example, when going to radiology, we alert the receiving department ahead of time).

The recent changes in the CF Foundation infection control guidelines were presented to the national CF community at the fall 2013 North American Cystic Fibrosis Conference. The updates are from those that were published in 2003. They are based on more recent clinical and research experiences. The goal of these guidelines is to minimize bacterial exposure from one CF patient to another. It is expected that all centers which are part of the Cystic Fibrosis Foundation Care Program adhere to the Foundation Guidelines.

Our team has reviewed the new guidelines, and as a result, we have decided we need to make some modifications to our existing infection control policies in order to be in compliance. Overall, the changes required to our policies are minimal, and it is our hope that we communicate these changes with you in this letter.

We strive to minimize any cross-infection risk to individuals with CF, and these revised measures will be implemented throughout our hospitals over the next few months. The major changes resulting from the new guidelines are as follows:

- **Masks:** Masks should be worn by all CF patients upon entering the hospital. Masks will be available at the front desk in the main lobby as well as the ambulatory clinic areas. Masks must be worn at all times in the hospital except when in an exam room or in a patient's hospital room.
- **Keeping distance from other patients with CF:** CF patients should maintain a minimum of 6 feet from other CF patients in all settings (except family members with CF) to reduce the risk of transmission. Previous guidelines recommended a 3 foot distance.
- **Attendance at events:** People with CF should avoid activities and risk factors known to be associated with transmission of CF pathogens. As a result, at our hospital CF Center and CF Foundation sponsored events, we can only allow one CF patient to attend. We encourage attendance by family members and participation by CF patients via webcasts.

As a reminder, medical evidence shows that all people with CF could carry bacteria or other germs that might be spread to others with CF in their lungs and sinuses. People with CF and those who do not have CF should clean their hands using soap and water or an alcohol-based hand sanitizer that is at least 60% alcohol.

If you have any questions about our infection control policies, including questions about any of the updates resulting from the new guidelines, please feel free to contact the CF center or speak with your CF clinical care team.



Valuable resources

LAURIE DOOLAN

My daughter just turned 20 and my son is 15 years old, so you might say that over the years we have become experts in cystic fibrosis and pretty much know all there is to know about managing the disease, right?

I mean, I try to keep up to date on research and treatment, schedule routine exams, enforce high calorie diets and snack eating, monitor that the kids are doing daily maintenance and taking all of their supplements and medications. I really didn't think there was much we didn't know about CF and its treatment. It is not always easy—as we all know, to take the time off of work and school for routine appointments, and three months seem to come up very quickly, but it is something we all must do. When it comes to CF Clinic visits, the tactic is get in and get out, avoid the unnecessary chatter, and try to get back on the road before the traffic. I will confess to being guilty of many eye rolls and sighs when it has been suggested that we see one of the specialists. I mean, we have been managing all of this for years, what more can they tell us? Well, I learned the hard way that maybe there is a thing or two to be learned. I have had to recently admit to myself that maybe we don't know it all. Maybe there is merit to the barrage of people whom they want us to see during routine CF clinic exams.

Years ago when my daughter was around 7, we were given the option of changing her physical therapy (PT) from manual to the Vest. She had really never been a fan of manual physical therapy and so the Vest seemed like it would be a great option for her. At the time, we opted to have our son continue doing manual PT with a therapist because that

was working well for him. So, they sent us the compressor (which was a huge machine back then!), and my daughter was fitted for Vest size. We were told which settings she should use on the machine, and that was it—we were on our way. She settled into a routine of doing the Vest after school and during the summer, and we even dragged that huge machine with us on vacations! For a half an hour every day (well, most days). Things with the Vest were working out so well for her that when my son's schedule started to get fuller, and it was becoming more difficult to fit in the daily therapist appointments, we decided to change him over to the Vest as well. Again, he was fitted for his own Vest, given settings, and we were told they could share the same compressor. We developed good routines and continued in our use of the Vest for years!

Recently, my daughter has had a steady decline in lung function, cultured some pretty bad macro bacteria and has a lobe of her lung that has significant damage. When it was suggested that we meet with Ann Gould from PT I, had my usual eye roll response. Really? What was she going to be able to tell us that we didn't already know? We had been doing daily therapy with the Vest for years and had even learned to use the new acapella during a recent hospitalization; we had this whole chest PT thing under control. Well, as I am sure you are probably guessing, it turns out maybe we didn't know it all, and there was much to be learned! We were scheduled for a half hour appointment with Ann before our regular appointment and, let me tell you, it was worth every moment of our time. It turns out that for all of this time—for years—we have not been using the Vest effectively. In the past, we had been taught how to use the machine, but my kids were never shown proper techniques: the breathing and

the huffing in between; the steps that produce results from the treatment. Right away, when my daughter started to do the steps as Ann showed her, she could feel a difference in the treatment. The appointment was so simple and yet, there was so much gotten from it. Before I left that day, I scheduled for my son to meet with Ann before his next appointment—and again, it was time so well spent.

As the parent of CF children, there can be a lot of guilt associated with making sure that they stay well and healthy and that I am on top of potential health problems and issues. I feel tremendously guilty and downright foolish that we have been using the Vest improperly for such a long period of time. When I think of the wasted time spent on a daily basis, I could scream! But the bottom line is that we can only do the best we can do and learn from our mistakes. I have certainly learned a valuable lesson from this situation and have developed a new outlook on how I will treat routine clinic visits in the future. I am sure that I will need to remind myself once in a while, but I am going to make an honest effort to try to use all of the valuable resources that are available through the CF Clinic and stay as long as I need to when I am there. We have recently met with the nutritionist and a gastroenterologist to discuss weight issues, and darned if we didn't learn some new tricks with these people as well! CF is a lifelong disease, and we need to remind ourselves that although we do know a lot, we may not have all the answers. If we are taking the time to drive into Boston, we might as well take advantage of any new insights we can be given. Traffic be damned!

Mother of two children with cystic fibrosis

Research

In 2013, over 1,300 patients participated in CF research studies across the country, more than in any previous year.

At Boston Children's, over 240 patients and their family members enrolled in observational studies, and 80 subjects participated in clinical research studies. We want to take this opportunity to thank these patients and families for their involvement in CF research. We would also like to thank all of the patients and families who expressed interest in participating in the Phase 3 Vertex trial for patients with two copies of DelF508. Enrollment in this trial completed two months ahead of schedule, and we look forward to results in the near future. Advances in CF care are not possible without your commitment to research. These individuals participated in the following studies:

- Vertex VX-770 (ivacaftor) for mutations R117H, G551D, and other gating mutations

- Vertex VX-809/770 for genotype DelF508
- Cayston and TOBI in Continuous Antibiotic Cycles
- KaloBios Pseudomonas Antibody
- Grifols Alpha 1HC inhaled anti-inflammatory agent
- EPIC observational study
- Pulmozyme study using ERapid nebulizer
- Newborn Observational Feeding Study
- Vertex VX-770 (ivacaftor) for 2–5-year olds
- Inhaled Vancomycin against MRSA
- N30 Pharmaceuticals Phase 1 trial of CFTR corrector
- Collections of blood, urine, sputum, and saliva for multiple studies

Boston Children's will be a part of a large number of CF research studies in 2014, and we are always looking for subjects who may be interested in learning more about clinical research. This year projects to be one of the most significant years ever in CF clinical research. We are currently enrolling or

anticipate beginning enrollment soon for subjects in the following studies:

- PTC Ataluren (PTC124®) for stop mutations
- Grifols Alpha1-HC inhaled anti-inflammatory drug
- Collections of blood, sputum, and urine for multiple studies
- Phase 2 study of IV Gallium for patients with Pseudomonas
- Phase 2 study of Novartis Potentiator for Class III
- Inhaled Hypertonic saline use in 2 to 5 year olds
- Study of Azithromycin and TOBI in patients .with first time Pseudomonas
- Two more studies that target the DelF508 mutation

This year saw the expanded approval of Kalydeco for several CF-causing gating mutations, as well as the enrollment and subsequent completion of the largest CF clinical trial(s) ever performed. The Vertex Phase 3 studies of VX-770/ VX/809 enrolled over 1,000 subjects from across the globe.

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



Boston Children's research team

A patient's perspective on exercise with CF

JASON HODGKINS

Patients with CF around the world are all too familiar with Airway Clearance Techniques (ACTs) and inhaled therapies to keep our lungs free of mucus.

I would know because I have cystic fibrosis and have been dealing with ACTs and inhaled medications for most of my 38 years. While all these amazing therapies have no doubt contributed to extending my life, my brother's life, and the lives of thousands of other people with CF, the one constant that I feel has contributed the most to my health is swimming.

My mom got me into swimming at the early age of 4 while we were living in Irvine, California. My mom knew that aerobic exercise was important but I don't think she really understood at that time how important, it would end up becoming. Swimming would become my sport and exercise throughout childhood and into my teenage years. When my family moved back to Massachusetts, I ended up joining the "Seagulls," a USA swim club located at Boston College, swimming every day except Sundays. Practice was grueling, and the time commitment was enormous, but the swimming benefited my health tremendously. I can't recall ever being hospitalized during my childhood. I eventually swam for my high school and was good enough to become team captain in my senior year.

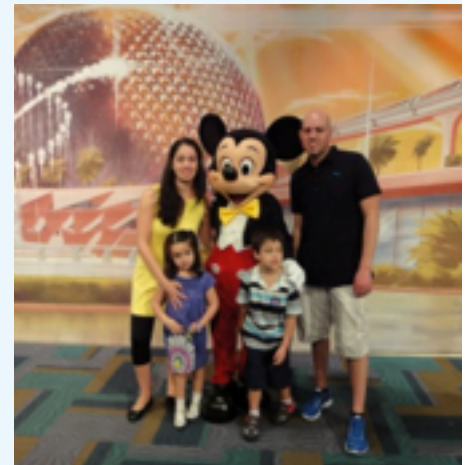
While my athletic career ended after high school, I have never stopped swimming. At 38, I'm still swimming at least 3 to 4 times a week. Yes, I've lost some lung capacity and endurance over the years. However, I'm still swimming about 40 to 50 laps depending on my health that day, and my lungs always

feel so free and powerful after a good swim—something everybody else would take for granted. But to someone with CF, the feeling of being able to take a deep breath and feel the fullness of air in my lungs is a motivating factor I've learned to appreciate.

The importance of aerobic exercise for people with CF is that swimming makes your lungs more powerful and efficient, according to Sharon Plowman and Denise Smith, authors of "Exercise Physiology for Health, Fitness and Performance." They explain that you must exert yourself when blowing out air under the water to overcome hydrostatic pressure, and that the process eventually improves lung strength. They also explain that breathing during all strokes except the backstroke requires you to pace taking in breaths, which results in deep breathing. You inhale fully after emptying your lungs and gradually increase lung capacity as a result. They also suggest that swimming's horizontal position maximizes your lung function potential.

Swimming has been and will continue to be a huge factor in my life; I hope to continue seeing the benefits of this wonderful exercise along with those from my normal ACTs and inhaled therapeutic regimens for a long time to come. I would strongly recommend swimming or any other endurance sport, including cycling, rowing and running to anyone with CF, young or old. Any endurance sport like swimming will be difficult at first, but the benefits you will feel will be immediate and may reduce the amount of time being sick or being hospitalized. Good luck and now get out and exercise!

Jason, 38, is married with twins and works full time.



CF updates and events

Navigating the Teen Years

This past fall Dr. Georgina Garcia, Judy Bond, SW, Lynne Helfand, SW and Kate Barnico, RN led a workshop for parents of teens and preteens with CF. The workshop was entitled "Navigating the Teen Years." There was a panel discussion led by a CF parent and an adult with CF. The class was well received by families.

Cystic Fibrosis Awareness Day May 2014

The Cystic Fibrosis Foundation hosted Cystic Fibrosis Awareness Day at the Massachusetts State House on May 1st. This event brought together Massachusetts Legislators and CF advocates, researchers, and caregivers. The event kicks off Cystic Fibrosis Awareness Month (May,) and the series of Great Strides the CF Foundation hosts each year to raise funding for CF research. The event was moderated by MassBio's President and CEO Bob Coughlin. Proclamations were awarded in recognition of Massachusetts CF Care Center Excellence.



Boston Children's Hospital receives recognition for Massachusetts CF Care Center Excellence

Hearing Screens

We recently started a new clinical initiative at the Cystic Fibrosis Center to include regular hearing tests as part of care for all children with CF. We have worked with the Audiology Department at the hospital and are recommending hearing screening at least every 4 years throughout childhood. Children and adults with CF are often treated with antibiotics that may affect hearing. We are therefore recommending hearing tests to monitor for any potential change in hearing. We are starting the process for our patients who are around 4 and 8 years old. Audiometric testing can be done on the same day as a regular CF clinic appointment. Our team will be working with you to schedule these appointments over the next year. These tests are done in the outpatient audiology clinic on the third floor of 333 Longwood Ave across the street from the main hospital building.



Great Strides 2014



Exercise

SHANE SOLAR-DOHERTY

Whenever I go in for a CF checkup, I reflect on two questions: Have I been consistent about my daily treatments, and have I followed a regular exercise regimen?

The first is usually a no-brainer, because if I'm not consistent about my treatments, I feel like I'm walking around with a bunch of cotton balls stuffed into my wind pipes. The second can at times feel a bit more arbitrary, though I know that exercise is just as vital for my health as my treatments. I have gone through periods when I didn't exercise very much at all and my health suffered, and other times when I exercised so much I started to identify with Superman and my health flourished. It took time and trials, and even some Internet surfing and YouTube watching, but I have come to an exercise routine that works well for me now.

The most important part about my exercise regimen is variety. There is no one exercise that is the absolute best for our health. They're all good. For that reason, I like to challenge myself and mix up my daily routine. I perform a variety of exercises from day to day, including running, biking, rowing, strength training, aerobic training and yoga. For a time I even saw a personal trainer who taught me different exercises for just working with body weight. The changing routine helps me to stay motivated, knowing that I'll be working different muscles each day. I try to get my heart rate up each time I exercise because I know that means my lungs will start working harder too. As I've found, performing different types of exercises has not only improved my lung function, but it has also strengthened my muscles, joints and bones for all different types of activities and stresses I place on my body day in and day out.

I make sure to feed my body well before and after exercise. A healthy diet is vital for keeping my lungs strong. I have found that eating a diet heavy in plant foods has worked best for me. Because I still need to keep my weight on, I make sure to eat foods that have high fat content, like nuts and seeds, healthy oils, and avocados. Every person with CF is different; for me, I have found that plant foods are much easier on my system to digest. Many plants contain naturally occurring digestive enzymes that aid in our digestion of them, and so my body can focus less on digesting food and more on improving my lung function. After I exercise, I usually drink a smoothie with kale, beets (great for lungs!), banana, berries, peanut or almond butter, coconut oil, chia seeds and almond milk. That's a drink that's tasty, replenishing and high in calories, and that makes me feel good after finishing.

For me, maintaining my health is all about consistent treatments, exercise, and nutrition. It's my trifecta. When I'm on my game with each of them, the cotton balls vanish and my health thrives.



Wyatt Mitchell and parents with Dr. Vo and Juliana Bailey, RD



Cystic Fibrosis Center clinic staff

A patient's perspective

** Please consult your CF Care Team before changing your health care regime.

Contact information

Appointments

617-355-1900, option 3

Live call hours

617-355-1900, option 4
8:30 to 11 a.m., Monday–Friday

Pulmonary function test

617-355-7510

Page operator

617-355-6369

Hospital main number

617-355-6000

It is recommended by the CF Foundation for CF patients to see a nutritionist, physical therapist and social worker yearly as needed. To schedule an appointment with one of these disciplines, call the scheduling center at 617-355-1900.

Changes to the phone line

The Health Care Question line (617-355-1900 option 4) now has “live call” hours from 8:30 to 11 a.m., Monday–Friday. Live call times are for you to use if you or your child is sick and may need to be seen. These live calls go directly to a nurse.

As a friendly reminder; the health care question line does not accept automatic refills from pharmacies. Parents / Patients / Pharmacists need to call and request refills. This will help prevent any inaccuracy in medications.

Thank you

The CF Center would like to thank the Nelson Family for contributions of toys, books and stickers to the pulmonary clinic. Patients and families have enjoyed the many donations. Thank you!

Medication and insurance

Are you having issues with receiving your medication and insurance coverage? Please call 617-355-4311 for questions and concerns.

Flu vaccines

REMINDER! Flu Vaccines are now available in Pulmonary Clinic.

Call to make an appointment: 617-355-1900



Cystic Fibrosis News Wire

Cystic Fibrosis Center
300 Longwood Avenue, Boston, MA 02115