CYSTIC FIBROSIS News Wire

FALL 2011

Welcome to the latest edition of the Children's Hospital Boston CF Center Newswire. We hope that the summer has been enjoyable and that you and your families have weathered the "back to school" frenzy that starts each fall.

This summer has been quite busy at the Children's Hospital Boston CF Center. We have welcomed three new pediatric pulmonary fellows, and our adult CF program has added another physician. Additionally, we have hired a new CF nutritionist. Our clinical research team has been very active conducting many different research studies, and our multi-disciplinary staff is now preparing for the annual North American Cystic Fibrosis Conference in November. Many members of our team will be leading workshops, presenting cases, or discussing their research at the conference.

In this newsletter, we are delighted to have more stories submitted by our families, in this case, families that have travelled to Boston from abroad to seek care at Children's Hospital Boston. We also have excerpted portions of an interview that was conducted several months ago by WBUR with an inspiring young woman who participated in a clinical trial at our CF Center. We hope you continue to find our newsletter a valuable source of information. If you have other ideas or suggestions for us, please do not hesitate to let us know.
Happy fall!

Gregory Sawicki, MD, MPH
Associate CF Program Director

Erica Sutherland's Great Strides team wearing t-shirts Erica designed for CHB/BWH Great Strides Team

SOCIAL WORK CORNER: STAYING CONNECTED

Several social networking and informational sites have become increasingly popular with patients who have CF and their family members. We recommend that patients under 18 years of age get their parents' permission before accessing these sites. We also encourage you to speak with your provider about any questions and/or issues that may arise after using these resources…and always remember that patients are not encouraged to have face to face contact with other patients and that if they do have contact, patients need to follow strict contact precautions guidelines.

We are excited to pass along the following list with a description of each site:

- **www.cff.org** The Cystic Fibrosis Foundation's website provides updates on cystic fibrosis treatments and research and provides information sheets about living with CF.
- **www.CFLiving.com** This site has an educational focus and is sponsored by Genentech.
- **www.cfvoice.com** This is an online community for children, adolescents and adults with CF, and caregivers. It has videos, articles, podcasts, and games and is described as "a place for motivation, inspiration, and connection to the CF community."
- **www.cffone.com** This site offers a social networking experience which is designed to promote treatment adherence in adolescents and young adults who have CF. It provides opportunities to have a personal profile, receive reminders for important CF and personal events, and learn about CF-related activities and research.
- **www.starlight.org** Starlight offers innovative media based programs to help individuals with CF enhance their quality of life. Starlight also invites families to join their Great Escapes program which sponsors special outings and events. The Great Escapes program staff take into account the need for implementation of infection control guidelines if more than one patient with CF attends an event.
- **www.TipsforCFParents.com** This site provides tips and resources for parents of children with CF and is written by a mother of two kids with CF who is also coauthor with Foster Cline, MD of *Parenting Children with Health Issues.*
- **www.cfrounttable.com** This online newsletter is topic focused and is written by adults who have CF.
- **www.cysticlife.org** CysticLife has been recommended by a number of adult patients with CF and their partners. It has informational and educational videos, and provides members with the opportunity to have interactive contact.

Enjoy…and let us know what you think of these sites!

OUR LITTLE ANGEL

Brenda Diaz, Mother

Doriangelic Marie Mejía Díaz

Nuestro ángel, Doriangelic Marie Mejía Díaz, nació un 22 de diciembre del 2008 para venir a hacer un cambio total en nuestras vidas y darnos mucha alegría. El 25 de febrero del 2009 dia de nuestro tercer aniversario queda recluída con una hemoglobina de 7 puntos, lo que al siguiente dia la lleva a la unidad pediátrica de intensivo en el Hospital San Jorge en Puerto Rico y una posterior transfusión de sangre que salvó su vida. Cuando la vimos abrazando la muerte comenzamos a preguntarnos Dios mío pero porque a nosotros? Le pedimos tanto y tanto a Dios que se apiado de nosotros y nos la devolvió a la Vida.

Es en Puerto Rico donde se hicieron infinidad de análisis de sangre que nos llevaron a concluir que fue una intolerancia a la leche lo que causó tal episodio.

Estuvo en monitoreo continuo con todas los especialistas hasta cumplir un año sin subir adecuadamente de peso y empeorando cada vez más su salud respiratoria. Cuando ya no pudimos mas con su estatus respiratorio decidimos cambiar de neumólogo y es entonces que nos recluyen por segunda vez sin aún conocer un diagnóstico preciso.

Aunque ya los médicos sospechaban de su condición, mamá y papá no querían aceptar. Continuaban nuestras preguntas, de porque nuestra beba? Esto NO nos puede ocurrir a nosotros? Mucha negación, entre otras...

Es el 9 de febrero 2010 es cuando nos reciben en el Centro Médico de Puerto Rico para hacer la prueba del sudor; confieso que fue la peor tarde de nuestra vida, aunque ya pasado un tiempo no creo que lo haya sido porque ese mismo dia recibimos un diagnostico; nuestra bebe era positiva para Fibrosis Quistica. Ahora si nuestras preguntas eran de mucho dolor y hasta pregunte a Dios porque no era un diagnóstico de cáncer u otra cosa que conociera y tal vez pudiese tener una cura o una solución? Desconocía mucho ya que en mi país no conocía nadie con tal condición.

Demás esta decir que aunque nuestro mundo se derrumbó de inmediato comenzamos los tratamientos sugeridos por su doctora y a seguir adelante!

Es en mayo del mismo año cuando llegamos al Boston Childrens Hospital para no tan solo transformar nuestra vida sino la de nuestro ángel. Comenzamos un tratamiento completo para eliminar la seudomona de sus pulmones, tratamiento con enzimas para procesar adecuadamente los alimentos y mejor que todo un adiestramiento para conocer su condición y manejarla en casa.

Al regresar a casa no tan solo tenía una niña que respiraba mejor, comenzaba a ganar peso y crecía dentro de lo esperado sino una niña FELiZ y muy juguetona.

Ahora le proveemos enzimas cada vez que come. Estamos al pendiente de todo lo que come y que tenga una dieta balanceada. Ofrecemos terapias respiratoria al menos dos veces al dia (pulmozyme y hyper – sal) con movimiento de secreciones. Al momento ya tenemos tres cultivos de garganta negativos para seudomonas por lo que se eliminó la terapia de tobramicina, GAD.

Ganamos gran confianza y albergamos una GRAN ESPERANZA en los tratamientos venideros.

Ahora vivimos bajo mejores medidas sanitarias para proteger la salud pulmonar de nuestra hija. Tenemos mas trabajo como familia porque hay que cumplir con el tratamiento no importa el cansancio ni los compromisos contraídos, pero vivimos mas confiados viéndola crecer con calidad de vida.

Ya no nos hacemos mas preguntas, solo agradecemos a Dios todo el tiempo por tantas y tantas bendiciones. Gracias Mi Dios por NUESTRO ANGEL...

ENGLISH VERSION:

Our angel, Doriangelic Marie Mejía Díaz, came into our lives on December 22, 2008, completely changing our lives and giving us a lot of happiness. On February 25, 2009 (day of our third anniversary) she was found to have a hemoglobin level of 7. The next day she was transferred to the Pediatric Intensive Unit at San Jorge Children's Hospital in Puerto Rico. A blood transfusion saved her life. When we saw her so sick, we began to ask God, "why us?" We asked God to save her life so we could take care of her.

She was continuously monitored until her first birthday. She was not gaining weight well and her pulmonary health was worse every day. When her pulmonary health worsened we decided to change our pulmonologist and she was admitted again without a diagnosis.

Her new pulmonologist spoke to us about the possibilies of a Cystic Fibrosis diagnosis. In February we took our baby to Centro Medico in PR to perform a sweat test. It was the worse day of our lives, but thinking back it was not, because that afternoon we received a diagnosis: our baby was positive for Cystic Fibrosis. Now our questions were of pure pain: I asked God why she did not have cancer, a condition that I know and that had a possible cure. We knew nothing about the condition and we did not know anybody in PR who had Cystic Fibrosis.

Our world had changed. It was a mess at the time, but we immediately began the suggested treatment and we had to move forward!

In May 2009, we arrived at Children's Hospital Boston our lives were tranformed. We were admitted for 13 days to eliminate pseudomonas from her lungs. We began a complete treatment with pancreatic enzymes so she could digest food adequately. But best of all, we knew more about the condition and how to manage her at home.

Now we provide her enzymes with every meal and snack. We are looking at how she eats, and that she eats what she needs. We give her pulmozyme and hyper-sal on a daily basis. We took TOBI and at this time we have had three negative cultures for pseudomonas, so Tobramycin was eliminated.

When we were back home in Puerto Rico, we had a baby who breathed better. Doriangelic began to gain weight adequately and grow up as a healthy baby and she is a happy, HAPPY baby.

We have gained a lot of confidence and hope during the upcoming treatments. We now live under better sanitary conditions to protect Doriangelic's pulmonary health. Of course we have more work because we have to do treatments no matter what!!! But we live better by seeing her grow with a higher quality of life.

Now we do not have more questions, we only thank God for everything. Thank God for our Angel!

BRENNA MCINERNEY

Hi, my name is Brenna McInerney, I'm sixteen years old and I have a nineteen month old little brother with Cystic Fibrosis named Nathaniel. He was diagnosed at nine days old and we have been making regular trips to Children's Hospital Boston ever since. The wonderful staff has made our trips bearable; this includes Dr. Sawicki, Judy Bond, Brandon Duque, Kate Barnico, and Erin the nutritionist. Our lives have changed a lot because of this diagnosis; we have learned not to take every day for granted. We have been very lucky so far as Nathaniel has been relatively healthy, aside from two bouts of pneumonia. However, worrying about enzymes, infection control, and trying to fatten him up have become our daily lives. I have learned how to take care of a child with CF. I have learned how to administer all of Nathaniel's medications such as enzymes, vitamins and zantac, as well as nebulizer treatments. Though he takes enzymes like a pro, the nebulizer is not so warmly received and it has come to the point where my mom and I have literally had to hold him down to give him the treatment. Physical therapy is also another tough issue because he won't sit still for it; he is always on the go! Through this diagnosis we have become involved with the Cystic Fibrosis Foundation and are currently raising money for our second year of participation in the Great Strides walk for Nathaniel's team. One of our fund-raisers is selling "cure CF" bracelets that I designed, made and sold myself. By having a child with Cystic Fibrosis come into my family I have learned so much, both about the disease and about life. Though this has been a hard road, and it has changed my life completely, I wouldn't trade it for the world. Nobody knows what the future holds, but we just take it day by day and hope for the best. I love Nathaniel so much and I'm so glad that he is in my life.

Michael 13, Brenna 16, Nathaniel 19 months, Olivia 9

MY MIRROR OF STRENGTH

By Callie Brochu

When you see strength what do you see? Do you see the big flexed arms and biceps, or the invisible scars that no one can see from afar, but up close, your eyes tell the whole story?

Strength doesn't come from the touch of a wand, you have to earn it. You have to have the chemicals of it streaming in your blood before you define yourself as "strong." Being strong shapes you as a person and you will find out that it has made my scars almost seem beautiful. It is a part of every word you say, every smile, move and decision you make.

I realized this at Children's Hospital, sometime in March, of 4th grade. I had just had severe surgery to my stomach and wasn't able to walk until sometime mid-week. When I was able to walk though, I took a look in the mirror. And behind all the tubes, dark under my eyes, grungy hair, and possible defeat hanging over my head, I found my own strength inside me. I had survived what some CF kids lose their life to, and that thought burned a scar in me so much deeper than I thought it would.

I went to sleep that night realizing that some of the best types of strong come from plane crashes, cancer, surgery and traumatic things that have imprinted us all. Those experiences make us carry our heads taller, pick up our feet, and get out the door. Being strong for someone makes you stronger in some way, because their pain is inflicted on you, too. I learned this when I was in the hospital actually, I had to be strong for my mom, I knew she needed me.

I believe that your strength will find you somehow and that it shows up at the most unexpected times. Strength has shaped me into being someone I never thought I would be… and I benefit from that.

Strength gives me the fire to fight and has formed me into someone who stands up when a crisis occurs, and doesn't stay sitting. It's like a suit of armor, keeping me ready and protected from some of life's harsh reality.

I didn't know how strong I was until strong was the only thing holding me together. I recognized that when the mirror showed me my strength… my mirror of strength.

NEWS FROM ICELAND

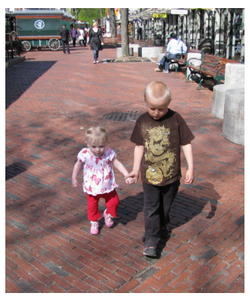
Frida Bjork Arnardottir

Our world collapsed on October 20th 2008. We were in Boston with our newborn daughter, who had undergone a cardiac surgery just a few days earlier. She was being transferred from the cardiac ICU to the floor unit and we, the parents, were happy that everything had been going according to plan. How wrong we were. We got the announcement that Maria had been diagnosed with Cystic Fibrosis. That is about all I can remember as I didn't have the faintest idea what Dr. Allan, Maria's cardiologist, was talking about. I had never heard of Cystic Fibrosis. I remember my husband listening to her and by the look on his face I could see the news wasn't good. When he explained the situation to me I broke down. How could it be that my beautiful baby, who already had a serious heart condition, had another horrible disease?

María Gunnarsdóttir age 2

A few minutes later we were brought over to 8 East where our communication with the CF team began. Dr. Christopher Hug came and spoke to us along with a colleague and explained what it all meant. I didn't really absorb the information at that point.

Everybody did a great job in enlightening us on the big task ahead. We got the feeling that Dr. Hug and Kate Barnico RN had taken us under their wings and they came by every day to answer our questions and just generally to see how we were doing. Every day they did their upmost to make us feel as well as possible. We got our enzyme feeding lessons and our PT lessons and some general lessons on how to keep her healthy and safe. Our most helpful lessons were the ones where we were constantly reminded that our daughter still could lead a fulfilling and meaningful life and do all the same things as other kids.



Living in Iceland, where Cystic Fibrosis is very rare, we hadn't heard of the disease before our trip to Boston. Only seven other children in Iceland have it. Even though we live in Iceland we always feel like we have direct access to Dr. Hug and Kate Barnico they are always ready to help us with whatever we need. Dr. Jónasson, our pediatrician in Iceland, also communicates with the CF team in Boston. Together they do their best so that Maria can lead as healthy a life as possible.

CLINICAL RESEARCH UPDATE

Erin Leone Thakkallapalli, MPH, CCRC, Program Co-Manager

Below are excerpts taken from Carey Goldberg's article "Special Report: New Cystic Fibrosis Drug Brings Gift of a Future," which appeared on May 6, 2011 in WBUR's CommonHealth Blog. Read the full article and watch videos at: http://commonhealth.wbur.org/2011/05/cystic-fibrosis.

In her entire 41 years, Roe had never been able to shovel snow. Or to run. Or to go a full winter without getting ill enough to need weeks of intravenous antibiotics.

She was born with cystic fibrosis. At birth, doctors told her parents that her life expectancy was age five. Along with school and play, her youth consisted of hospital beds and piles of pills and hours each day of inhalation therapy.

Medical treatments that continually improved in small steps, from new antibiotics to improved enzymes, kept her alive. But she remained a person without a future, told at each life stage not to expect the next. Her husband planned for retirement. She did not.

Now, because of a still-experimental drug called VX-770, made by Cambridge-based Vertex Pharmaceuticals, that has changed Roe clearly has a spirit as sparkly-bright as a Roman candle. When she came to speak to the cystic fibrosis team at Children's Hospital Boston recently, she wore zebra-print stilettos that made the same fun-wild statement as the many shades of red in her hair. But she is openly bewildered by the new possibility of living out a full lifespan.

"Now I'm like, holy cow, I might be 80!" she said. "Maybe I should start using some face cream!" It's almost like she's a different person: "I still have cystic fibrosis but I can do things, I can live my life."

Roe does not use the word "cure." No one does when they talk about VX-770. More cause for optimism? Yes. But a cure? No.

Roe still takes antibiotics and does hours of inhalation therapy every day. She doesn't dare stop. But "this is life-changing," she said. "I'm very, very careful, I'm trying not to get excited, but I really am, at the same time."

This is two stories, intertwined. One is Roe's life with cystic fibrosis and then on VX-770. The other is a tale of amazing science—and a gamble of hundreds of millions of dollars that hit the medical jackpot. Researchers tell it with a hint of disbelieving awe in their voices: For once, nature played no tricks. For once, everything came together just as it was supposed to, from theory to test-tube to human patients. The story of VX-770 reflects a new scientific era, or at least, aspects of how this new scientific era we're in is supposed to work.

"It has dramatically changed this disease from one of despair to one of hope and hopefulness," said Dr. Robert Beall, head of the Cystic Fibrosis Foundation, which was instrumental in the drug's development.

If there is a useful moral to this rare tale of scientific success, it may be the kindergarten maxim: Work together. The Cystic Fibrosis Foundation, founded in 1955, is held up by many as a model disease-fighting group that bridges between researchers, doctors, patients and donors in perpetual quest for a cure. Without the foundation, everyone seems to agree, there would be no VX-770.

The foundation is still far from all it aims to achieve. Though VX-770 is nearing the end of clinical trials, its long-term effects remain unknown, and it only appears to help 4% of people with cystic fibrosis, those with a specific mutation known as G551D.

But still, there is good reason to revel in this particular moment in biomedical history. In key ways, scientists really cracked this disease. That knowledge has been translated into one drug that has seen resounding initial success in clinical trials, and it looks likely to lead still farther. And Roe is wondering if she should go back to school.

When Roe was born, in 1970, cystic fibrosis had been identified as a distinct disease for more than a generation, but little was understood about the mucus problems that affected multiple organs in patients' bodies. As Roe grew up, she remained resolutely upbeat, but she doesn't pretend that life with cystic fibrosis was easy. She missed a lot of school, fell behind on schoolwork and had trouble keeping friends—she was "out of sight, out of mind." She missed prom. And "Dating—that was a joy! Dating is tough enough without, 'Oh, by the way, I have this mucus issue? Let's go out!'"

Through the 1980s, the Cystic Fibrosis Foundation was ramping up its efforts, setting up a network of special care centers and another network of research centers around the country. In 1989, a team led by Francis Collins—who now heads the behemoth National Institutes of Health—pinpointed the cystic fibrosis gene, and the protein it makes. Known as CFTR, it is involved in transporting sodium and chloride ions across the cell membrane.

It was a moment of great uplift in cystic fibrosis circles. A cure seemed conceivably within reach.

Roe participated in various studies over the years—for antibiotics, inhalers and supplements—but "It was always like 'Ooh, this is the next best thing" but "once you got the med, it was just another drug to help me maintain."

You can particularly understand how Roe was not especially keen on research given that through her twenties, the trendiest direction in cystic fibrosis research was gene therapy, the idea that you could replace the defective cystic fibrosis gene with a good one. Long story short, it fizzled.

There were other advances: a lung drug developed specifically for cystic fibrosis; aerosolized antibiotics that could be inhaled. But no game-changers. And slowly but surely, as with most cystic fibrosis patients, Roe's lungs were getting worse. She worked full-time in the cosmetics industry but she had to miss work often; she coughed blood; she came to have trouble keeping her blood sugar stable—cystic fibrosis often brings with it diabetes.

Meanwhile, Bob Beall of the Cystic Fibrosis Foundation reached a conviction in 1998 that had him burning up the phone lines. Persuaded that the way forward for developing a cystic fibrosis drug was through high-throughput screening—testing tens of thousands of compounds for effects on CFTR—he called one drug company after another in hopes of enlisting them in the effort. Aurora Biosciences of San Diego—which was later acquired by Vertex—called back.

"We then committed $40 million to do high-throughput screening to see if we could find one or two molecules that could open up the sodium channels," Bob recalls. "People thought I was crazy." (Though, he hastens to add, "my board was 100% behind me.")

Eric Olson, who oversees much of the cystic fibrosis work at Vertex, explains about high-throughput screening: "You always find something," some compounds with some effects. But "they may not be potent enough, they might be toxic—they're never the drug, they're a starting point, what we call a scaffold, that you turn over to the medicinal chemists," for tinkering.

His team figured they were actually after two molecules for two different defects involved in different cystic fibrosis mutations. Work on the two ran neck and neck, but one molecule pulled ahead, even though only a small minority of patients had that mutation. "We had to get something into the clinic," Eric said.

VX-770 went into its Phase I clinical trial in humans—a small, brief initial trial checking mainly for safety—in 2006. In early 2007, the larger Phase 2 opened. The buzz started small. At a cystic fibrosis meeting, a nurse involved in the trial came up to Eric and said, "I know I'm not supposed to tell you anything, but I'm so excited! This drug is doing something! It could be the placebo effect, but I've known these patients for years, and this drug is doing something."

'I wanted to believe it," Eric said, "but I didn't want to go down this path of false hope."

Word spread. People were saying things like "That patient feels better than he has in years." But of course it was all anecdotal until the official findings came in. When Eric's colleagues, Drs. Claudia Ordonez and Bob Kauffman, called him in to a Vertex office to present the first Phase 2 data, it all became real.

"I was in tears," Eric said, visibly moved even by the recollection. "What was most dramatic was the drop in sweat chloride, because it really meant it was working."

No one had ever done this before. VX-770 had actually lowered patients' sweat chloride score, and not by a little. Some scores were cut in half. "When it actually happens in real people—it's amazing."

When the results were presented in the spring of 2008 at a national cystic fibrosis meeting, the crowd was electrified. Eric remembers people saying, "This is the most exciting thing we've seen in my 30 years of working on cystic fibrosis."

Roe wasn't excited, though—not yet. She passed up her first opportunity to join the Phase 2 VX-770 clinical trial; it just wasn't the right time for her. Finally, in the fall of 2009, she decided to try the little blue pill at the center of so much buzz. Or rather, she enrolled in a double-blind trial, so she didn't actually know whether she was on VX-770 or placebo.

At first, she thought maybe she was just having a good week, or month. Then, she started to bring up more mucus. "Actually, it felt like a flash flood," she said. "I would cough and it would just come flying out of me, not to be too gross. It was just everywhere. It was much thicker than it used to be. It was constant, all day long, and I thought, 'Oh, crap, I'm getting sick,' but I never actually got sick."

"And as time went on, my amount of mucus decreased, and my cough decreased. Then I started noticing bigger and bigger changes. I was sleeping. I was able to maintain my blood sugars. I could basically eat and not have to worry. I no longer needed any medication for constipation. And the biggest thing for me was, that year was the first year in my entire life that I never got sick."

Roe has a port in her chest for intravenous antibiotics. She hasn't used it since she started the trial in 2009. And more: before she started on the trial, she had a "huge mucus plug" in her chest so bad that she was considering surgery to remove part of her lung; now it appears to be gone.

As she spoke to the Children's Hospital cystic fibrosis team, she mentioned that her FEV, a measure of her lung function, had been 62%. Now it's 81%.

"Oh my God," exclaimed Nancy Shotola, who runs the pulmonary function lab.

Roe still has a bit of a cough, but it's more like throat-clearing than the old hacking. She still wheezes a bit, with asthma. But she never coughs blood anymore. She can clean her house. She runs two miles every other day. She feels so normal that sometimes, amazingly enough, she almost forgets to take her blue pill.

Roe is still taking VX-770 as part of a clinical trial, but now it's "open-label," and she knows she's on the drug, not placebo. Vertex is planning to apply to the FDA later this year for permission to market it. Somehow, she says, until a doctor can pull out a pad and write her a prescription for whatever VX-770 ends up being called, "it's not completely real."

Vertex is not saying at this point how much the drug will cost—though she has asked. It is expected to be expensive.

Vertex is testing VX-770 in children, with excellent results. In February, it announced the data from a second Phase 3 study on VX-770, this one in children as young as six—also predictably excellent. And it's testing a combination of VX-770 and a different drug, VX-809, a "corrector" aimed at bringing CFTR to the cell surface instead of just unlocking the gate, in adults with a far more common mutation. A second "corrector" drug, also aimed at helping the 90% of patients who have the more common mutations, is also being developed.

All good news. But it brings more "a sense of responsibility than elation," Eric Olson said. "Now that you know the answer, but you're only getting to 4%, and nobody else has anything in the clinic for the rest of these patients, it's a big responsibility. We've got to move faster, harder. It's almost like you've got the golden ring, and you know what it can do, but then you've got to figure out the way to do it."

It's a painful topic for Roe, the other 96%. Already, she's a senior citizen by cystic fibrosis standards. A friend recently had a double lung transplant. To friends with cystic fibrosis, and even to family, she doesn't tend to talk much about how VX-770 has changed her life.

But she's so naturally open that you only have to ask to get a glimpse of the huge emotional dislocation she's now navigating. She is reimagining her life. It's the opposite of the typical mid-life crisis in which people suddenly see death looming. Suddenly, she sees a whole unexpected second half of her life on the horizon.

"Hello," she said, "I'm here! Now what?"

On her wrist is a tattooed reminder, in graceful cursive, "breathe."

She may need it now as much as ever—though for different reasons.

ADULT CF CENTER UPDATES

Ahmet Uluer DO Adult CF Program Director

The adult cystic fibrosis program is excited to announce new additions to our team of care providers. As the number of adults with CF increases at our CF center, we have increased the number of CF clinicians. We would like to announce the arrival of Dr. Manuela Cernadas, a first class pulmonologist. She will be joining us in the adult CF clinic at Children's Hospital Boston (CHB), as well as Brigham and Women's Hospital (BWH). Dr. Carolyn Donovan will now be providing care in both the inpatient and outpatient setting. Dr. Craig Gerard will continue to care for patients in both the inpatient and outpatient setting. Leah Frain, family nurse practitioner (FNP), has been a part of our program already for some time now. She has increased her hours to 4 days a week and will continue the expert care our NP's provide. Dr. Ahmet Uluer, Neeli Sudin, FNP and other members of the BWH pulmonary division will continue in their current roles. Our multidisciplinary team of clinicians, physical therapists, nutritionists, social workers, respiratory therapists, and others, continue to work with you. Their names can be found on our website at https://www.childnrenshospital.org

Leah Frain NP, Ahmet Uluer DO, Carolyn Donovan MD, Craig Gerard MD, Simona Rits, Anne Gould PT, Neeli Sudin NP, Beatrice Duvert, Robin Welcher RD

We are excited about our involvement in the CF Foundation sponsored Learning and Leadership Collaborative, specifically the Adult Quality Improvement (AQI2) Initiative, with emphasis on improving respiratory and nutrition care. We are very fortunate to have many energetic volunteers on our team that will ensure our success, including Laura Alvino, a patient at our center, and Francine Healey, a parent of two of our patients. Other members (in alphabetical order) include Isabel Bailey, Carolyn Donovan, Beatrice Duvert, Leah Frain, Anne Gould, Paul Ricard, Simona Rits, Neeli Sudin, Ahmet Uluer and Robin Welcher. One of our endeavors includes a new exercise program with support from the Vertuccio family and aptly called the Steven J. Vertuccio Wellness Program. Our goal will be to perform annual evaluation of exercise capacity, as well as before and after an exacerbation that requires hospitalization, and will include an exercise prescription for home. The program will feature equipment produced by SCI-FIT, a company specializing in scientific solutions for patient fitness. Our exercise program will also take advantage of an additional feature called Fit-Key, which allows the clinician to download and review data collected from patients as they exercise. This will allow our physical therapists to show our patients the progress they are making and allow for improved continuity of care during an individual's subsequent admissions and follow-up in the outpatient setting. The data will be stored on an electronic 'key' for review by health care professionals and when the key is inserted into a machine; the program resumes that individual's preferred settings.

We have sent out a patient survey to better meet your needs in the outpatient setting. This survey is very important to us as we investigate ways to make your experience in clinic a more meaningful and efficient one. Our goal is to receive a completed survey from at least 80% of patients so please return this as soon as you can.

Another project we are working on is to increase your access to a nutritionist and make this interaction more personalized to meet your needs. We have implemented an algorithm for determining patients who are at nutritional risk. Our inpatient nutritionist completed a mentoring program through the CFF and we are sending the outpatient nutritionist to take advantage of a similar program. Our other projects include ways of improving your experience in clinic by introducing an electronic version of the pre-clinic form you are asked to fill out when you first arrive to clinic.

PARENT FAMILY ADVISORY COMMITTEE (PFAC)

If you are interested in becoming an active member please email cfevents @childrens.harvard.edu or contact Judy Bond, SW.

EMAIL

To receive updates from the CF center via email go to cfevents @ childrens.harvard.edu and add your address and name.

UPCOMING LOCAL CF FOUNDATION EVENTS:

10/01/11 Aptalis CF Cycle For Life ®
11/18/11 Uncork the Cure
For more information:
http://www.cff.org/Chapters/mass-ri

SEASONAL FLU VACCINE

THE CF CENTER IS KEEPING RECORD OF CF PATIENTS RECEIVING THE FLU VACCINE:

Email or call: cfevents@childrens.harvard.edu or 857-218-4625

Provide patient's name and where (primary care, Children's or other) vaccine was received.

Cystic Fibrosis Center at Children's Hospital Boston and Brigham and Women's Hospital "Cystic Fibrosis Night" will be held Spring 2012.

JULIANNA BAILEY, RD, LDN

I am thrilled to be joining the team at the Cystic Fibrosis Center of Children's Hospital Boston as a Registered Dietitian! I earned my Bachelor of Science in Nutrition and Food Science at Auburn University in Auburn, AL and completed my Dietetic Internship at the University of Alabama at Birmingham in 2010. During my Dietetic Internship, I completed a specialization in pulmonary nutrition which sparked my interest in working with patients with CF. While finishing my Master of Science in Clinical Nutrition, I was able to complete a Nutrition Traineeship with the UAB Pediatric Pulmonary Center. I worked as a clinical nutritionist in the Pulmonary Division of Children's Hospital of Alabama. While I was able to work with patients with a variety of pulmonary disorders, my concentration during my traineeship was in nutrition for Cystic Fibrosis and I completed my thesis research on Vitamin D supplementation in children with CF and vitamin D deficiency.

I am from Huntsville, AL and new to the Boston area. I love it here so far and feel very fortunate to have this opportunity to continue working to benefit children with CF! I am excited to begin my career with the CF center at Children's Hosptial Boston and look forward to working with everyone!

CYSTIC FIBROSIS News Wire

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