

Heart to Heart

Volume 7, Number 2 • Fall 2010 • Winter 2011

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Second International PEDIATRIC CARDIOMYOPATHY CONFERENCE Takes Place

CCF's Second International Conference on Cardiomyopathy in Children, co-sponsored by the National Heart, Lung and Blood Institute (NHLBI) and the University of Miami Miller School of Medicine, was held May 13-14 in Bethesda, Md. The scientific workshop was enthusiastically received by the research and medical community and called attention to critical issues related to cardiomyopathy in children. Additional meeting sponsors included the Myocarditis Foundation and GeneDX.

60 of the top clinicians and researchers from the U.S., United Kingdom, Canada and Australia convened to identify key research

areas, share best practices, and review new research data and findings on pediatric cardiomyopathy. A wide range of medical disciplines was present, including cardiologists, geneticists, molecular biologists and epidemiologists. Representatives from the NHLBI, National Cancer Institute, and the National Institute of Child Health and Development also participated in the exchange of ideas and information.

"Cardiomyopathies result in some of the worst pediatric cardiology outcomes, as nearly 40 percent of the children who present with symptomatic cardiomyopathy receive a heart transplant or die within the first two years after diagnosis," said

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CCF RAISES AWARENESS of SUDDEN CARDIAC ARREST on Capital Hill



Sudden Cardiac Arrest Coalition Members

In a powerful demonstration of collective standing, CCF along with more than 40 organizations representing the Sudden Cardiac Arrest Coalition (SCAC) gathered on Capitol Hill on October 5 to raise awareness of sudden cardiac arrest and

highlight the importance of cardiopulmonary resuscitation (CPR) and automatic external defibrillators (AED) in saving lives.

In a briefing held on the 50th anniversary of CPR, coalition members issued a call to action for people of all ages to learn how to recognize sudden cardiac arrest, learn CPR and get familiar with AEDs.

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Children's
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A Cause for Today... A Cure for Tomorrow

The *Heart to Heart* newsletter is published biannually by the Children's Cardiomyopathy Foundation (CCF), a national organization dedicated to saving lives and improving the quality of life for children with cardiomyopathy. CCF's mission is to accelerate the search for a cure by supporting research, educating physicians and patients, and increasing awareness and advocacy related to the needs of affected children and their families. CCF is a publicly supported tax-exempt organization as described under section 501(c)(3) of the Internal Revenue Service.

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From Lisa Yue, CCF Founder & President

In October, I traveled to Washington D.C. as part of the **Sudden Cardiac Arrest Coalition**, to raise awareness of sudden cardiac arrest (pg. 1). During the trip, I exchanged a few words with a fellow passenger who inquired about the nature of my trip. I told her about CCF, the families we help and our involvement in the Sudden Cardiac Arrest Coalition. She had never heard of cardiomyopathy before and didn't realize that so many deaths resulted from sudden cardiac arrest (more than 290,000).

These brief encounters happen often, and I always wish there was more time to educate people on cardiomyopathy and the work CCF does. If I had a copy of this newsletter, I would have shared with her how our **new school packet** responds to the challenges that school age kids with cardiomyopathy face (pg. 10). Or how **Dr. Carolyn Ho's genetic research** has the potential to prevent the disease in its early stages from manifesting (pg. 4).

If I could have a heart-to-heart conversation with all those I meet daily, I could share the hope behind the words on these pages – a hope of finding a cure and the belief that kids with cardiomyopathy will eventually have an active and healthy life. In this issue we interviewed **Ally Smith**, the young "bionic bride" who sustained her life with a heart assist device after experiencing heart failure from cardiomyopathy. Her story (pg. 9) is a wonderful example of how hope still exists for those living with this disease.

As we approach year-end, let us keep hope alive and do our part to raise public awareness of cardiomyopathy. Please support our "**Heart-to-Heart**" holiday appeal and together, we can give cardiomyopathy and CCF the attention it deserves.



Time for a HEART-TO-HEART



"There is so much I want to do, but my heart won't let me. It doesn't work like it should."

The Children's Cardiomyopathy Foundation is working hard to find the cause and cure for cardiomyopathy. But we need your help.

Please give today to support a better tomorrow for children living with this chronic heart disease.

An online donation can be made at childrenscardiomyopathy.org (click on "make a donation")

MYOCARDIAL FIBROSIS as an Early Manifestation of Hypertrophic Cardiomyopathy

Most patients with hypertrophic cardiomyopathy (HCM) develop more scar tissue in their hearts.

This is known as fibrosis and is abnormal because the healthy heart should not have scar tissue. This increased amount of scar tissue may be part of the reason why people with HCM have problems with heart failure or irregular, sometimes dangerous, heart rhythms. At this point, doctors do not know what causes this scar tissue to form. In a study by **Carolyn Ho, M.D.** of **Brigham's Women's Hospital** in Boston an intriguing question is raised: Is myocardial fibrosis (scar tissue) in HCM a late result of chronic disease, or can it begin early in the disease because of gene mutations, and perhaps even drive the development of HCM?

In her findings published in the August 5 issue of the **New England Journal of Medicine**, Dr. Ho states that levels of C-terminal propeptide of type I procollagen (PICP), a byproduct released when collagen is made by the cells of the body, were significantly higher in both

individuals with HCM and individuals who were HCM mutation carriers without left ventricular hypertrophy, compared to individuals who did not carry the HCM mutation. In this study, PICP was used as a blood serum biomarker or indicator for the formation of scar tissue in the heart. Of the measured biomarkers, PICP was the only biomarker to show any difference between the mutation carriers and non-carriers.

The increased levels of PICP, triggered by the mutation, suggest that more collagen is being made in the hearts of people with sarcomere mutations than in those without mutations. This seems to be happening even in young people whose hearts look normal and who have not yet been diagnosed with HCM. Increased formation of collagen may lead to scar tissue depositing in the heart. "These findings are interesting because they tell us that the development of fibrosis might play a role in actually driving development of the disease, rather than it being simply a reaction to or reflection of the development of overt disease

later in life," states Dr. Ho.

The study looked at three groups of adult and pediatric patients: group one had sarcomere mutations and clinical symptoms of HCM, group two had mutations but no left ventricular hypertrophy and group three was a control group with no HCM mutation. The research team used several serum biomarkers of collagen metabolism in addition to echocardiography and cardiac magnetic resonance imaging (MRI) to evaluate the differences in the three patient groups.

What Dr. Ho and her research team found was that patients with sarcomere gene mutations (groups one and two) had increased blood levels of the biomarker PICP. This suggests that their hearts make more collagen than subjects without sarcomere mutations and this leads to the abnormal formation of scar tissue in the heart.

These observations offer new insight into the way HCM develops. "It also raises the very interesting possibility that using drugs to block the development of fibrosis early on in these gene-mutation carriers without hypertrophy may help to delay HCM from developing," said Dr. Ho.

Second International Pediatric Cardiomyopathy Conference Takes Place continued from page 1

Conference Chair and CCF Medical Advisor **Steven Lipshultz, M.D.** "This conference is a critical step in bringing together the best in the field to understand the molecular and genetic issues in cardiomyopathy and to identify the most critical and promising areas for clinical research efforts for this disease," continued Dr. Lipshultz.

The two-day conference was organized into six moderated sessions centered on various topics of interest. Panel moderators included:

- **Genetic Issues:** Jeff Towbin, M.D.; Wendy Chung, M.D., Ph.D.
- **Molecular Mechanisms:** Steve Webber, M.B.Ch.B.; Daniel Bernstein, M.D.
- **Biomarkers in Pediatric Cardiomyopathy:** Steve Lipshultz, M.D.;

Michael Burch, M.B.Ch.B.

- **Hypertrophic, Restrictive and Left Ventricular Noncompaction Cardiomyopathy:** Steve Colan, M.D.; Robert Weintraub, M.B.B.S.
- **Myocarditis:** Leslie Cooper, M.D.; Charles Canter, M.D.
- **Medical & Transplant Therapy:** Daphne Hsu, M.D.; Robert Shaddy, M.D.

The conference also included five young investigators who presented their research findings for group critique.

A new family panel was introduced this year in which attendees had the opportunity to hear from parents of affected children. Five parents from the D.C. and Maryland area shared their experiences of navigating the medical system during

their child's evaluation and diagnosis, and discussed their challenges of raising a child with a chronic illness. They also offered suggestions on which patient resources and support services are most helpful and what physicians could do to further assist parents after a child's diagnosis.

Similar to CCF's first conference in 2007, manuscripts from this year's conference will be featured in three issues of **Progress in Pediatric Cardiology** in 2011. Remarkably, the 2007 issues resulted in the most downloaded articles in the 20-year history of the scientific journal. Plans are also in place for additional working group sessions to focus on discussed research priorities and to develop action plans for collaborative studies aimed at improving diagnostic methods and optimizing care for children with cardiomyopathy.

The **POWER** of genetics in

UNDERSTANDING and TREATING Hypertrophic Cardiomyopathy

Genetic discoveries have allowed remarkable advances in understanding inherited heart disease, such as hypertrophic cardiomyopathy (HCM). Over the past 20 years, researchers have determined that, in most cases, HCM is caused by changes (mutations) in genes that make up the molecular motor of the heart, called the sarcomere. Although most people who have inherited a sarcomere gene mutation will eventually develop HCM, traditional findings such as left ventricular hypertrophy are often not present early in life.

Genetic testing allows for early identification of family members who carry their family's HCM gene mutation and are therefore at risk for developing the disease, but at a time before their hearts show any obvious changes. Studying people with disease causing mutations before they show symptoms of the disease allows researchers to better understand how mutations lead to the development of HCM and the onset of symptoms. This information is crucial to develop innovative treatments that will hopefully one day be able to slow or even prevent HCM from developing.

Recently **HCM♥Net**, was established with funding from the **National Heart, Lung and Blood Institute** (NHLBI) to further research in this area. HCM♥Net is a multi-center collaborative network of dedicated HCM centers around the country, which will work together to advance understanding of HCM and develop new treatment strategies. **Dr. Carolyn Ho**, medical director of the hypertrophic cardiomyopathy clinic at **Brigham's Women's Hospital** in Boston is the lead investigator of HCM♥Net. She has been focusing on this line of research for several years. Dr. Ho's previous research highlighted that sarcomere mutations cause subtle changes in the heart wall before the onset of

increased thickness that characterizes HCM. The heart appears to have difficulty relaxing and be more prone to developing scar tissue even when it looks normal. "Although it is remarkable that we can do genetic testing to identify the exact cause of HCM in a family, we really want to put that information to practical work. Inheriting the mutation is like lighting the fuse. We would like to be able to lengthen or, better yet, cut the fuse before HCM develops. Understanding the steps leading from the mutation to HCM provides an opportunity to do just this," said Dr. Ho.

A major focus of HCM♥Net will be to study gene mutation carriers early in life, before HCM is diagnosed. "The characteristics that differ between the preclinical mutation carriers and their healthy relatives without mutations provide key clues about how HCM develops," said Dr. Ho. "These are the first steps on a long but important road. We hope to use the information and infrastructure we are establishing to actually start testing new treatments within the next five years. It will take a lot of time and work, but by working together — families, patients, scientists in the laboratory and doctors — we can transform the way we care for people with HCM."

The HCM♥Net study is currently in its recruitment phase and investigators are looking for HCM families with HCM to participate. All family members may be eligible for participation, including those with HCM, individuals who carry a sarcomere gene mutation but have not yet developed left ventricular hypertrophy, and healthy family members who do not carry the mutation. The goal is to recruit about 180 patients between the ages of 5-40 years old. If genetic testing has not yet been performed, this may be available free-of-charge for eligible families.

HCM♥Net is a multi-center collaborative network of dedicated HCM centers around the country, which will work together to advance understanding of HCM and develop new treatment strategies.

The study involves a single visit to one of twelve HCM♥Net centers. Participants will have their blood drawn and undergo exercise testing on a treadmill, ultrasound testing (echocardiogram) and a cardiac MRI. Completing all elements of the study will take approximately six hours.

HCM♥Net network sites include: Brigham and Women's Hospital, Boston, Mass.; Children's Hospital, Boston, Mass.; Cleveland Clinic Foundation, Cleveland, Ohio; Cincinnati Children's Hospital, Cincinnati, Ohio; University of Chicago, Chicago, Ill.; University of Colorado, Denver, Colo.; Johns Hopkins University, Baltimore, Md.; University of Michigan, Ann Arbor, Mich.; St. Luke's Roosevelt Hospital, New York, N.Y.; Stanford University, Palo Alto, Calif.; Vanderbilt University, Nashville, Tenn.; Washington University, St. Louis, Mo.

For more information on this study or to learn how to enroll in the study, please contact, Carolyn Ho, M.D. at 617-732-5685, cho@partners.org or Allison Cirino, CGC, 617-732-7921, acirino@partners.org.

Eighth Annual CCF Golf Classic a HUGE SUCCESS

On July 19, CCF sponsors, donors and friends enjoyed a wonderful day of good weather, great golf, and a memorable dinner and silent auction at the historic Montclair Golf Club in northern New Jersey. As in the past, the event boasted a sold-out crowd of 192 attendees from top name law firms, asset management firms, hedge funds and other financial service companies who helped raise \$405,390, surpassing last year's total of \$333,746. Net proceeds from this year's



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event will be earmarked for CCF's research and education fund.

In the evening CCF family member **David Fisch** of **Vanderbilt Appraisals** spoke movingly of his son Nate's diagnosis of hypertrophic cardiomyopathy and the invaluable support he found through CCF.

Prizes for the winning foursomes went to: **Al Lhota, Dan Powers, Rob Ambriano** (First prize); **Kevin Kavanagh, David Pucciarello, Jon Jachman, Frank Brosens** (Second prize); **John McCormick, Mike Dowdell, Nick Argiriou, Patrick Lanigan** (Third prize); **Tim Milton, Jim Joyce, Wendy Sacks, Russ Diminni** (Fourth prize).



Warming up on the driving range



Top photo: Attendees enjoying dinner

Bottom photo: Nicole & David Fisch with CCF founders Eddie & Lisa Yu

The prize for closest to the pin went to **Adam Shane** and **Andrew Finkelstein**. The prize for longest drive went to **Dan Powers** and **Mike McBride**. The raffle prize winner was **Pat Leahy**.

2010 Event Sponsors: Sunil Aggarwal; Dan Allen & Family; Anchorage Advisors; Angelo, Gordon & Co.; Barclays Capital; Richard Barrera & Purnima Puri; Chris Berry & Matt Lambert; Jay Bharadwa; Steven Bleier; Brigade Capital Management; Tom Boyce; Benji Cheung; Cantor Fitzgerald; Dustin Cappelletto; Chris Chang; Sheree Chiou & Jason Kirschner; Pierre & Robin Chung; Eric Cole; Costa Family; Credit Suisse; Davidson Kempner Capital Management; Deutsche Bank Securities; Lucas Detor; Robert & Karen Dishner; Euro Brokers High Yield; Fisch & Mansdorf Family; Robert Frahm; Fried, Frank, Harris, Shriver & Jacobson; Eric Friel; Rob Gallivan & Family; Michael Garrity; Gleacher & Co Securities; Sachin Goel; Goldman, Sachs & Co; Mike Henderlong & Family; Hunton & Williams; ICAP High Yield; David Johnson; Katten Muchin Rosenman; George Khouri; Kolatch Family Foundation; Clint Kollar; Kramer Levin Naftalis & Frankel; Daniel & Lauren Krueger; Zachary Lewis; Si Lund & Family; Mackay Shields; Mandel, Katz & Brosnan; Jim Martin & Heidi Hullinger; McDermott Will & Emery; Justin McEvily; Drew McKnight; McLaughlin Family; Morgan Stanley; Mitch Petrick; Gil Nathan; Jared & Carrie Nussbaum; Dan Ornstein & Family; RBC Capital Markets; Richards Kibbe & Orbe; Steve Rosen; Edgar Sabounghi; Ian & Mackenzie Sandler; Adam & Heather Savarese; Senator Investment Group; Chaney Sheffield; Taconic Capital Advisors; Vanderbilt Appraisal Company; Leigh & Esther Walzer; Chris Warren; Eddie Yu & Family.

Save the Date!

3rd Annual

All-In For A Cure No-Limit Texas Hold 'Em Tournament

**Wednesday, February 9, 2011
at Crimson, New York City**

Join CCF for an
exciting evening of charity poker!

Try your luck at
winning the grand prize:
entry to the
**World Series of
Poker Main Event.**

For more information,
please contact Sheila Gibbons at
866-808-CURE (2873) ext. 902.

More event photos are online under "News & Events/Golf Classic."

FAMILY FUNDRAISERS

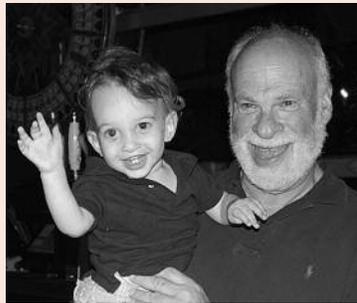
Furthering Our Mission

Recording Artist Zack Weber Sings for CCF

On September 15, a group of students from **Fontbonne University** in St. Louis, Mo. hosted a concert featuring fellow alum and up-and-coming recording artist **Zack Weber**. The event attracted more than 150 people and was held at The Label, a lounge popular with both students and locals. More than \$2,500 was raised in honor of Zack's two nieces, **Bryn Tracy** who has dilated cardiomyopathy and **Ava Tracy** who lost her battle with cardiomyopathy at six years old. "It was an honor doing this for CCF," said benefit coordinator and student **John Masidonsk**. "Especially after hearing **Chanda Tracy**, Bryn and Ava's mother, speak at the event. It was very touching."

Fisch Family Slam Dunk

Nicole and David Fisch really had something to celebrate when their son **Nate**, who was diagnosed with hypertrophic cardiomyopathy at three months, stabilized and turned one in July. The Fischs asked relatives and friends to donate to CCF in lieu of birthday gifts for Nate's party. As a result, more than \$8,000 was gifted in honor of Nate. At the birthday party, held at Carnival in New York City, guests were given a chance to dunk Nate's father, David, in a dunking tank with a donation of \$5 per ball. An additional \$600 was raised from this fun game, and Nicole's father, **Mike Mansdorf**, matched the amount.



Nate Fisch with grandfather Mike Mansdorf

Straight From the Heart Cardio-a-Thon

Jodie Sanchetta and **Dan Hornby** of the **Straight from the Heart**



Straight from the Heart Foundation members

Foundation, held its Second Annual "Cardio-A-Thon" on May 23 at the Verona Community Center, N.J. The exercise-oriented event was held in memory of Jodie's two friends, **Karl Weber** and **Rob Diess**, who lost their lives to cardiomyopathy. Jodie, a personal trainer, used her

incredible energy and skills for physical training to raise more than \$8,000 for CCF. 2010 event sponsors included **Fleet Feet**

Sports, Adrenaline Training Studio, Mary Kay Cosmetics and **Villani Chiropractic Center**.

Take CCF Out to the Ballgame

The Metro New Jersey Chapter of the **Appraisal Institute**, held a day at the TD Bank Ballpark in Bridgewater, NJ, for the Somerset Patriots game on May 23, which raised \$3,350.

Victor and Patti DiSanto, long-time CCF supporters, along with **Debra Miller** organized the event. The day included fun activities for kids and a chance to walk out on the field to meet the players. Victor and Patti's son, **Joey**, was diagnosed with dilated cardiomyopathy and had a heart transplant in 2006. This is the third year that the DiSanto family and the Appraisal Institute have partnered to fundraise for CCF.



Victor DiSanto & Debra Miller

Lenny Skates with Me

On April 10, **Caroline Parisi** and **Hockey**

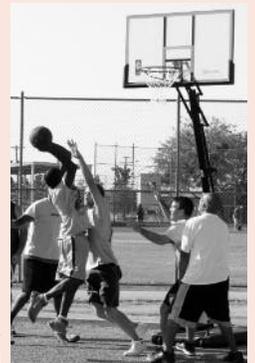


Hockey North America team

North America (HNA) held a hockey game in honor of Caroline's husband who had hypertrophic cardiomyopathy and died of sudden cardiac arrest at the age of 36. Friends and family came out for the second year to honor **Lenny Parisi**, who played for HNA for many years. The Wilmette Centennial Ice Arena in Illinois provided free rental for skating after the game.

Hoop Dreams

Casey and Heather Riley's son, **Casen**, passed away unexpectedly from hypertrophic cardiomyopathy at the age of six months. To celebrate what would have been Casen's first birthday on August 31, Casey and Heather hosted the "**Casen Riley 3-on-3 Basketball Tournament for Pediatric Cardiomyopathy**" in Abilene, TX. The tournament was a huge success with eight teams and many spectators. The outdoor event raised \$3,000 for CCF, and the Rileys also sold food, t-shirts, CCF curebands and raffle items. "It was a great day of friendly competition to not only honor the life of our sweet baby boy, but to also build awareness of cardiomyopathy," said Heather.



3-on-3 Riley Basketball

More Family Fundraisers...



Grace Callahan

Amazing Grace Doll Furniture

Inspired by his daughter, CCF family member, **Doug Callahan** turned his woodworking hobby into a creative way to raise money for CCF. Doug's 7-year-old daughter **Grace**, who was diagnosed with dilated cardiomyopathy and had a heart transplant in April, wanted to have a place to tuck in her American Girl dolls at night. They worked together to design a

wooden bed, and now Doug's handcrafted doll furniture is being sold online with net proceeds going to CCF. Grace's grandmother makes all of the mattresses and pillows by hand.

Since he started this summer, Doug has sold 13 beds and plans to add an old-fashioned school desk and armoire in the future. "It's been a fun way to raise some money for cardio-myopathy research and help families who face this difficult disease," Doug says. To learn more about



Amazing Grace wooden doll bed

Doug's wooden doll furniture, visit his site at sites.google.com/site/amazinggracewoodworking/. The price of a single bed is \$45.00 and a bunk bed is \$85.00 plus shipping (choice of painted or stained finish).

- 11-year-old **Griffin Ngai**, of Cedar Hill School, Basking Ridge, N.J., ran 22 laps in a fundraising challenge in May and chose CCF as the beneficiary in honor of his cousin, CCF family member, **Christopher Piccininni**. Christopher was diagnosed with cardiomyopathy at nine weeks and received a heart transplant at six months old.
- For the second year in a row, 6th grader, **Macie Bridge** of Groton, Mass., and former student of CCF family member **Scott Middlemiss**, raised more than \$400 asking family and friends to donate to CCF in lieu of birthday presents to honor Scott's son who has left ventricular non-compaction cardiomyopathy.

CCF Raises Awareness of Sudden Cardiac Arrest on Capital Hill continued from page 1

More than 100 people including representatives from the offices of Senators **John Kerry** and **Roger Wicker** attended the Washington D.C. briefing.

Sudden cardiac arrest (SCA) is the leading cause of death in the U.S. claiming more than 295,000 lives annually. While CPR can double the chance of surviving SCA, only one in four victims receive CPR from a bystander. The Coalition asked Congress to commit to three agenda items: join the **Congressional Heart and Stroke Coalition** to promote sudden cardiac arrest awareness, encourage state education departments to require all high school graduates to be trained in CPR and AED use, and fund research efforts that allow for consistent and accurate data collection across the country on sudden cardiac arrest events.

Keynote speaker **Lance Becker, M.D.**, one of the country's leading SCA response experts and a professor of emergency medicine at the **University of Pennsylvania**, emphasized that more research in the field of cardiac arrest resuscitation and more uniform outcomes and performance measurements are needed to increase SCA survival rates. "It's been said before that you can't improve what you don't measure and the same applies to sudden cardiac arrest," said Dr. Becker. "Until we bring the needed resources, research and national standards for data collection to the forefront, we won't know for sure what works, what doesn't and why.

Following Dr. Becker's appeal to policy makers, **Brian Buck**, a 30-year-old SCA survivor, told his story about how proper SCA response saved his life. He shared details of the day when his heart stopped suddenly and how his colleague, **Cheryl Victoria**, responded to the emergency with CPR and an AED. "There is not a day that goes by when I don't feel incredibly blessed to be alive and grateful for what Cheryl did to save my life," said Buck at the briefing. "My hope is that many more people would be trained in the use of AEDs and CPR, and survival stories like mine would become much more prevalent."

The Coalition also recognized three journalists and a congresswoman for their outstanding achievement in SCA awareness. **Robert Langreth** and **Matthew Herper**, both senior editors at **Forbes**, were recognized for their extensive reporting on SCA. **David Epstein**, a staff writer for **Sports Illustrated**, was recognized for writing a 2007 cover story about hypertrophic cardiomyopathy, the leading cause for sudden cardiac arrest in young athletes. Congresswoman **Betty Sutton** (D-Ohio) was recognized for her sponsorship of the **Josh Miller HEARTS Act**, which authorized the Department of Education to provide funding to schools for purchasing AEDs.

Read about SCA survivor Brian Buck's story on page 8.



Brian Buck and family

BRIAN BUCK: HCM Genetic Carrier and SCA Survivor

CCF family member, Brian Buck, was invited to Capital Hill by the Sudden Cardiac Arrest Coalition to tell his story of surviving a sudden cardiac arrest. Below are excerpts from his speech.

Hello, my name is **Brian Buck**, and I am 30 years old. I'm married to **Julie** and have two children, **Aubrey** and **Malakai**. Two and a half years ago a heart murmur was detected in Aubrey at her six-month well-baby check-up, and we soon learned she had a heart disease called hypertrophic cardiomyopathy or HCM. I also found out I had HCM as a result of Aubrey's diagnosis, but I was told that the disease should not affect me and I could continue life as normal.

So I continued with my active lifestyle and on June 16 of this year I went on lunch break from my engineering desk job at **Conoco Phillips** in Texas to play soccer next to our employee fitness center. As I was walked away from the field after scoring a goal, I collapsed on the sideline next to 22 fellow soccer players – none of whom realized I was in cardiac arrest. Our fitness coordinator, **Cheryl Victoria**, was the first emergency responder on the scene. Realizing I did not have a pulse, she hooked me up to the automatic external defibrillator (AED) immediately. The device shocked me and then Cheryl began cardiopulmonary resuscitation (CPR) with the assistance of another co-worker. After four rounds of compressions and breaths, my heart regained its pulse.

The ambulance arrived within 10 minutes of my collapse. At the hospital specializing in cardiac care, my body was cooled to 91 degrees through cryogenic therapy to help preserve brain function. I awoke after the sedation period just in time for my wedding anniversary and Father's Day. I learned I had a cardiac arrest and would soon receive an internal cardiac defibrillator.

I feel like it was a miracle I had my arrest in a place where people had the equipment and training to save my life, and I'm here today telling my story to help make sure other victims get the same second chance at life. My hope is that the survivor story will become much more prevalent.

I have since had a complete recovery from the arrest. The only change is I am learning the best approach to activity levels to make sure another emergency event doesn't occur. For the rest of my family, since my arrest we have learned that my son is gene positive for HCM in addition to my daughter. Overall, my family is doing well, and Aubrey has been relatively healthy.

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Brian with his rescuer Cheryl Victoria

CCF and the CURE THE PROCESS Campaign

For several months, CCF has been involved in the “**Cure the Process**” Campaign, which focuses on creating new science-driven public policy that will improve the **Food and Drug Administration (FDA)** regulatory process related to the development of rare disease therapies. More than 150 partnering organizations, including CCF, recently placed calls to **Senator Kohl (D-WI)** and **Senator Brownback (R-KS)** of the **Agriculture, Rural Development and FDA Appropriations Subcommittee** to request a \$10 million programmatic appropriation for the FDA to create a new drug review division for rare diseases. Thanks in part to the campaign, a hearing on rare disease regulatory issues was held.

As a result, the FDA's **Rare Disease Committee** has been tasked with submitting a report to Congress in March of 2011 on the recommended regulation changes for rare disease drug development. Additionally, a bill supporting the creation of new clinical study designs and analysis for rare disease treatments was submitted. The bill also includes a funding increase for the orphan product development grant and the creation of an associate director for rare diseases position at the **Center for Drug Evaluation and Research**. The next step is for **The Senate Appropriations Committee** to review the bill. More information about Cure the Process is available at curetheprocess.org.

A SECOND LEASE ON LIFE

Allyssa Smith, the “Bionic Bride”



At 21, **Allyssa (Ally) Smith** was fighting for her life while being treated for advanced heart failure caused by dilated cardiomyopathy. Her one wish was to make it to her dream wedding and marry her college love **Mike Babineaux**. This summer, her story of how she resumed her life with the assistance of an implanted heart

pump was featured on the **Today show, Fox News** and **People Magazine**. Recently, Ally shared her remarkable story with CCF.

In October 2006, Ally was a vibrant 18-year-old college freshman, an avid athlete and the picture of good health. But then she started feeling fatigued, suffering dizzy spells and fainting during athletic events. To her dismay she found out she had dilated cardiomyopathy, most likely caused by a viral infection. In the years that followed, she and her cardiologist **Roberta Bogaev, M.D.**, of the **Texas Heart Institute** tried many treatments in the hopes of improving her heart function and keeping her off the heart transplant list. However her condition deteriorated in 2009, and it suddenly looked like she would not survive to see her wedding day.

Time was running out for Ally, who had not been successful in gaining a donor heart. Dr. Bogaev suggested one final option—implanting a left ventricular assistance device (LVAD) to do the work of her faulty heart. The revolutionary device, which some have dubbed as a “bionic heart,” sustains heart failure patients until a donor heart becomes available. The bigger hope is that the heart assist device or heart pump will give the heart enough time to rest and recover so that once the heart pump is removed a heart transplant is no longer necessary. The miniaturized mechanical device would be inserted in Ally's chest cavity and be connected to her heart with an external long-lasting battery pack worn by Ally as a shoulder bag. The device would essentially take over the pumping function of the weakened heart muscle, allowing blood to circulate throughout the body more effectively. In April 2009, with the support of her family and fiancé, Ally underwent the eight-hour surgery to have the **Heart Mate II®** assist device implanted. This life-saving heart device has helped some 4,000 patients like Ally sustain their lives.

Since the procedure, Ally says, “I have felt better than I had for the three years before the [implant]. Granted, it took my body a while to acclimate to the LVAD, and everyone is different, but I definitely could tell a difference after a few months. Physically, I cannot feel the LVAD in my heart. There are parts of the machine that are pushed up against my ribs and stomach that I can see and feel, but as far as when I move, can I feel it? No.”

The only impediment that the LVAD presents is that Ally must avoid contact with water (the tubing leading to the battery pack can not be submerged). While she was a former competitive

swimmer, she now stays away from pools and the ocean. She also bathes with a special waterproof bag on her shower door and stays inside on heavily rainy days. Aside from this, Ally can do everything else a normal person does. “I live off batteries during the day, and at night I hook up to a portable battery unit that connects to the wall outlet,” she explains. “I am electric, but surprisingly that does not affect my life as much as one would think.”

Her medical team in Houston praise Ally's courage and poise. In July, Ally finally realized her dream of walking down the aisle in her gown and leather cowboy boots. She is now continuing with her education and adjusting to life with her new husband. She also has taken on a new role as an advocator for heart disease. Ally devotes a great deal of time to educating the public on the impact of heart disease and the importance of a timely and accurate diagnosis. She has shared her story with local and national media, as well as with American Heart Association chapters throughout southern Texas. She sees the need to increase awareness of cardiomyopathy and to demonstrate that heart disease can affect a healthy, athletic college student just as it can affect someone who is much older and less fit.

Ally continues to volunteer at Texas Heart Institute to talk to pre-operative and post-operative LVAD patients and their families about her experience with the heart pump. Her time spent with patients has been so rewarding that she is contemplating becoming a surgical nurse.

The advice she most often gives to families of people living with cardiomyopathy is, “Be the support system that someone with cardiomyopathy needs. Do not ignore them especially if they are saying that something doesn't seem right — having a support group is the most important thing a patient with heart failure needs.” Her best advice to her fellow young patients is, “Do not ignore symptoms even if they seem small. Talk to your doctor about everything, and if you are not sure about something or don't like the answer that you get, demand an explanation so that you can better understand what's happening to you. Be your own advocate! And keep living your life. Life is too precious to think it is over just because you are sick.”

Ally's prognosis looks good. Dr. Bogaev believes Ally could survive five to 10 years with her HeartMate, buying her additional time for a heart transplant if needed. In January 2011, Ally will find out if she can safely remove her LVAD and whether the combined efforts of the heart pump and cardiac medications have given her heart enough of a reprieve to heal itself and function on its own. There are signs her heart is recovering and has miraculously begun to augment the work the pump is doing. “Over time we've seen that her heart is taking on more of the [pump's] work,” says Dr. Bogaev. If her heart can function normally on its own after the assist device is removed, Ally will really have a reason to celebrate. The first thing Ally plans to do upon full recovery is “to do a bomb shell into the pool!” To us at CCF, Ally is an inspiring example of how to live a full life in spite of a chronic heart condition.

Expert Q&A:

Working with the School System to Ensure a Quality Education



CCF's newest educational resource, **Ensuring a Good Learning Environment: A Cardiomyopathy Resource Kit for School Personnel and Parents** was developed to educate school staff about pediatric cardiomyopathy and to guide families in working with the school to create a safe and supportive learning environment for their child.

CCF recently asked three parent members who assisted in the development of the school resource kit to share their experiences of working effectively with their child's school. Kate Middlemiss, a kindergarten teacher; Scott Middlemiss, an assistant principal (parents to Joseph, 3, HCM); and Terri Dermeyer (mother to Michaela, 9, HCM) share their tips.

What should parents bring to the school meeting?

Kate & Scott: Parents should bring a number of different documents including:

- Medical summary sheet from cardiologist and/or other doctors
- List of medications with dosage, time given, possible side effects
- Contact information for all medical professionals on the child's team
- Letter describing parent concerns
- All evaluation reports from school, hospitals and outside sources

In some cases, parents should bring other professionals to the meeting. Any early intervention therapists or case manager who worked with your child can attend to speak to the more specific needs around speech, occupational therapy (OT) or physical therapy (PT). A healthcare professional from your

child's medical team can attend to help bridge the medical/school gap. If the parents are not comfortable with special education laws, they may choose to bring an advocate to help with the process.

How can the school help inform classmates about the disease?

Terri: If you are able to, go into the school and talk to the class about the diagnosis. Show pictures of the heart to the other students. I make sure to tell them specific examples of how they can help. I say, "If you see Michaela lying on the ground and she doesn't get up, have one person stay with her and another go get an adult. Even if you have to run in the hallway you won't get in trouble."

Kate & Scott: Classmates usually embrace the idea of differences and will help to support and welcome children with different educational and medical backgrounds when the teacher creates a healthy classroom environment. Social skill lessons should be geared toward the acceptance of all differences without isolating specific children. Classroom activities can be developed to promote the sharing of personal differences (for example, "Star of the Day" or "I am special" units).

You may want to visit the classroom

yourself or invite the school nurse or other medical personnel to discuss the disease. If the child feels comfortable, have him/her speak about the disease and how it impacts his/her life. You don't have to share all the details, but the more friends and classmates are aware, the more they can do to help. Remember, the teacher will not be able to watch your child every minute, so it will be good for classmates to be aware of warning signs and symptoms as well.

How does the school keep parents updated, such as when a staff member notices a symptom, there is an illness going around, or an education plan must be modified?

Terri: The teacher usually calls me or sends a note home if there is an illness going around the school. The teacher calls if several kids are sick in my son's classroom as well (he does not have cardiomyopathy but he can bring home the illness to my daughter).

Kate & Scott: Be sure to provide the school with numerous contact numbers so they can easily reach you. Another idea is to start a daily or weekly communication journal between the staff and home. Finally, request phone updates (daily, weekly, monthly) or

periodic meetings with all staff who come in contact with your child.

What can parents do to be more actively involved in their child's school?

Kate & Scott: Find out about the school's "open door" policy and your availability to visit, observe or volunteer in the classroom.

Why is it important to keep written records of contacts and conversations parents have had with school staff?

Terri: When you keep good records it can help when a question comes up about how a situation should be handled. It protects you as well as your child and the school. It can also help down the road with a different school year. You can refer to your records and say, "This is what so-and-so did in this situation."

What is the most important tip to working effectively with school staff you can give to other parents?

Terri: Let the staff know how much you appreciate what they are doing for your child. Listen to what they are telling you, because sometimes they come up with really important things.

Kate & Scott: Your child becomes your teacher's "kid" the seven hours or so he/she is at school. A teacher who has a student with a serious medical condition will most likely feel anxious or nervous, especially if they are unfamiliar with the child or the condition. Parents have to always keep in mind that their child is one of 25 (or more), and the teacher is dealing with other medical and educational issues. So, this is stressful for the teacher, as his/her ultimate goal is to keep all students safe.

Parents can validate the stress and nervousness while clearly outlining

parental expectations in order to create a parent-teacher team atmosphere from the start. Introduce yourself to the principal and assistant principals and make sure they are fully aware of the situation. Create an emergency plan with the staff that correlates with the student's schedule and placement throughout the building. All staff members working with the child should be CPR trained and fully educated on the child's Individual Medical Plan.

Emergency plans should be made available for substitute teachers and specialists too.

Daily academic pressures are difficult for any child. What are some tips for parents on reducing school stress for their child?

Kate & Scott: Ask about the availability of support from a guidance counselor.

Guidance counselors may have friendship groups or lunch groups that meet with other children dealing with medical issues. This can help your child know there are other kids in his/her school dealing with some similar challenges.

What People are Saying about CCF's School Resource Kit

"I used the format letter in the packet and that got the ball rolling at school. The school was immediately attentive to Ryan and his condition and our situation like never before! I was able to meet with the principal, Ryan's 6th grade teachers and also his physical education teacher. They all know now and how to respond if something were to happen. Thanks so much for helping our children live their lives safely."

Faith Settles

Mom to Ryan (11, HCM) and Chance (9)

"It's a great resource for nurses and it alleviates a lot of the parents' burden of 'educating' the school about the child's diagnosis and special needs. It also decreases the stress and anxiety of the school personnel in receiving these children."

Jenny Nova, Pediatric Heart Failure & Transplant Coordinator
Montefiore Children's Hospital

"I have spent the better part of my day trying to find educational material for my daughters' school that are easy-to-understand without too many medical terms. With two children who have HCM, I need to inform several staff members at the school and the school aftercare every year. The aftercare program even had a meeting with all of their staff members using the school packet as a learning tool."

Kristin Burgos

Mom to Isabelle (11, HCM) and Emily (7, HCM)

Ensuring a Good Learning Environment: A Cardiomyopathy Resource Kit

is viewable online at childrenscardiomyopathy.org (click on Support Services/Educational Resources). To request a hard copy, please contact Kella Boyer at 866-808-CURE ext. 905 or kboyer@childrenscardiomyopathy.org.

HearttoHeart

Volume 7, Number 2 • Fall 2010 • Winter 2011

The Newsletter of the Children's
Cardiomyopathy Foundation

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CCF Forum Guest Q&A Sessions

- **Parenting Teens with Cardiomyopathy or a Heart Transplant**
Miriam Kaufman, BSN, M.D., FRCPC,
Adolescent Health Specialist
Hospital for Sick Children
November 8 – 15, 2010
- **Medications and Treatments for Pediatric Cardiomyopathy**
Daphne Hsu, M.D.
Chief of Pediatric Cardiology
Children's Hospital at Montefiore
December 6 – 13, 2010
- **Feeding Tubes and GI Issues**
Elizabeth Gleghorn, M.D.
Director of Pediatric Gastroenterology
and Nutrition
Children's Hospital Oakland
January 10 – 17, 2011



Local Support Groups

- **North Carolina**
Duke University Medical Center
Durham, NC
January 27, 2011 at 6:30 p.m. – 8:00 p.m.
February 24, 2011 at 6:30 p.m. – 8:00 p.m.
March 24, 2011 at 6:30 p.m. – 8:00 p.m.
April 28, 2011 at 6:30 p.m. – 8:00 p.m.
May 26, 2011 at 6:30 p.m. – 8:00 p.m.
Room 4902, McGovern-Davidson
Children's Health Center
- **Massachusetts**
Brigham and Women's Hospital
Boston, MA
November 20, 2010 at 10 a.m. – noon
Shapiro Building,
6th floor family conference room



Phone Support Group

- **Pediatric Heart Transplant**
For members with a child who has received or may receive a heart transplant
November 11, 2010 at 8:00 p.m. – 9:00 p.m.
Eastern Standard Time
- **New Member or New Diagnosis**
For members who are new to CCF or who have children that were recently diagnosed
January 22, 2011 at 1:00 p.m. – 2:00 p.m.
Eastern Standard Time
- **Parents of Teens with Cardiomyopathy**
For parents of a child age 12 or older with cardiomyopathy
November 10, 2010 at 8:00 p.m. – 9:00 p.m.
Eastern Standard Time