

Heart to Heart

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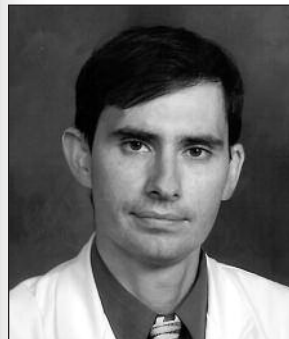
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CCF Announces 2011 RESEARCH GRANT Recipients

The Children's Cardiomyopathy Foundation (CCF) has awarded \$150,000 in seed funding to three new research studies on pediatric cardiomyopathy for 2011. These studies will focus on the areas

of pediatric heart transplantation and genetic mutations related to dilated cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy type 5 in children.



Carmelo Milano, M.D.



Jill Tardiff, M.D., Ph. D.



Kathleen Hodgkinson, Ph.D.

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CCF Funds MULTI-CENTER HCM Study

CCF is initiating a new multi-center research study, *Exome Sequencing to Identify Novel Causes of Infantile Hypertrophic Cardiomyopathy*, led by Wendy Chung, M.D., Ph.D. of Columbia University Medical Center. Other participating centers include Pediatric Cardiomyopathy Registry sites such as Cincinnati Children's Hospital, Children's Hospital at Montefiore in New York, Children's Hospital Boston and Holtz's Children's Hospital in Miami.

CCF will provide \$100,000 to fund the study aimed at identifying new genetic mutations that cause hypertrophic cardiomyopathy (HCM) in children under one year of age. Families with more than one affected child without an identifiable genetic cause are of

particular interest because these families are most likely to have an underlying novel genetic mutation.

For infants with cardiomyopathy, it is particularly challenging to determine the cause of the disease in a family because far less is known of cardiomyopathy in this age group compared to older children and adults. Infants with hypertrophic cardiomyopathy tend to have a worse outcome, but it is still unclear what genetic factors contribute to its severity in the young. "Because of their poor prognosis, determining the genetic cause for cardiomyopathy in infants becomes even more important," says CCF

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A Cause for Today... A Cure for Tomorrow

Children's
Cardiomyopathy
Foundation

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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All submissions and correspondence regarding the newsletter or Foundation should be directed to:

Children's Cardiomyopathy Foundation

P.O. Box 547
Tenafly, NJ 07670
Tel: 866.808.CURE
Fax: 201.227.7016

info@childrenscardiomyopathy.org
www.childrenscardiomyopathy.org

The Children's Cardiomyopathy Foundation, including all parties to or associated with *Heart to Heart* will not be held responsible for any actions readers take based on their interpretation of articles from this newsletter. As always, readers are encouraged to discuss medical evaluations and treatments with their own physicians.

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From Lisa Yue, CCF Founder & President

Dear Families,

I had the privilege of participating in the recent "*Genetic Diseases of Children: Advancing Research and Care Conference*," hosted by the New York State Department of Health, and I was particularly moved by presentations from **Fred Modell**, co-founder and president of the **Jeffrey Modell Foundation** and **Jim Kelly**, Hall of Fame NFL quarterback for the Buffalo Bills and co-founder of the **Hunter's Hope Foundation**.

Both shared their journeys of finding hope in the midst of suffering after losing their sons to rare genetic diseases and how from their own experiences they formed foundations dedicated to helping others in need. Fred Modell's speech focused on how to build upon the unexpected events in your life, and Jim Kelly spoke about how sports taught him to never give up even when fighting for his son. Those who know my story of losing two children to cardiomyopathy can understand why their words and actions are so inspiring to me.

We all know there is no more passionate advocate than a parent with an affected child. In this issue we highlight another group of exceptional individuals – dedicated scientists and physicians, impassioned families, and generous supporters and volunteers who are all helping to call attention to pediatric cardiomyopathy and taking the steps to defeat this disease. I hope you enjoy reading about these ardent advocates and the special ways they are making a difference for children with cardiomyopathy.



Lisa Yue

Research is Essential

We need your help in raising critical dollars for studies that will lead to eventual cures for cardiomyopathy in children. Watch your mail for CCF's spring appeal mailing or make an online donation today to support research for a healthier tomorrow.

An online donation can be made at childrenscardiomyopathy.org by clicking "Make a Donation."

A heartfelt thank you to all our 2010 sponsors and donors!

Read CCF's 2010 Annual Report online under "About CCF/Financials."



2011 RESEARCH GRANT Recipients

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EXPANDING THE DONOR HEART POOL

Carmelo Milano, M.D.

associate professor of surgery at Duke University

Cardiac transplantation remains the most effective treatment for children with cardiomyopathy progressing to end-stage heart failure. However, the shortage of available donor hearts continues to be a primary limitation. While mechanical heart pumps are being utilized by adults to extend the life of their failing heart, this therapeutic option is still under development for the pediatric population. Therefore, children with advanced heart failure awaiting heart transplant experience a three-fold increase in death rate relative to adults.

Currently, pediatric heart transplants are only using hearts from deceased donors that are brain dead but continue to have cardiac function. An alternative organ donor exists in the form of an individual who has suffered severe brain injury but is not declared brain dead. Hearts from these donors are removed only after life support is withdrawn, the heart has stopped beating and the donor formally declared dead. Although organs from these types of "donation after cardiac death" (DCD) are being utilized in non-cardiac organ transplants (liver, kidney), there is concern that DCD hearts are not useable because of tissue damage that occurs from the lack of oxygen during cardiac arrest.

Dr. Milano's study focuses on expanding the pool of available donor hearts for children with advanced heart failure by proving that "donation after cardiac death" (DCD) hearts can be safely utilized for pediatric transplants. Dr. Milano believes there are two factors in favor of using DCD hearts for pediatric transplantation. One advantage is that DCD donors tend to be younger than BD donors, and the second advantage is that hearts from

BD donors who have experienced cardiac arrest have been previously transplanted successfully.

Dr. Milano will attempt to establish the degree of injury that the human heart can sustain during the DCD or cardiac arrest process and still be suitable for transplantation. He also will look at the amount of recovery achieved by his proposed preservation process. The ultimate goal is to develop a method to halt or reverse the heart injury sustained during cardiac death and develop an accurate method to predict the functionality of the DCD heart prior to transplantation. Dr. Milano will examine human hearts from adult and pediatric DCD donors that have been treated with a special preservation solution designed to minimize heart injury and compare them to human hearts from BD donors that have been immersed in conventional storage solution. Comparison of the data from the different donor hearts will help develop the parameters for determining heart quality.

CCF family member, Linsey Rippey, feels lucky her family only had to wait 18 hours for a donor heart for her daughter, Madison, who was transplanted in August 2009. "It was very daunting knowing we could be waiting months for a heart for Madi," says Linsey who sees the significance of Dr. Milano's study. "I have been there, and while my daughter is one of the lucky ones, far too many children are not as lucky."

According to Dr. Milano, half of the heart transplants required in the near future will be for cardiomyopathy patients. "Currently, the donor pool is inadequate with few suitable pediatric donors," he says. "The long term goal of this study is that someday a child with cardiomyopathy will be able to use DCD donor hearts for transplant."

STUDYING GENE MUTATIONS THAT CAUSE SEVERE PEDIATRIC DCM

Jill Tardiff, M.D., Ph.D.

associate professor of physiology and biophysics at Albert Einstein College of Medicine

Dr. Tardiff will use her grant award to clarify the molecular mechanisms that underlie early onset dilated cardiomyopathy (DCM) and to determine whether they may be generalized to other human genes that cause DCM. Specifically, she wants to understand how a single amino acid substitution in Tropomyosin, a sarcomeric protein that makes up the structure of the heart muscle, causes a more severe disease in children than in adults. This novel mutation (D230N) was recently shown to cause a complex cardiomyopathy that presents in early childhood with heart

CLINICIANS & SCIENTISTS

Request for Research Applications on Pediatric Cardiomyopathy

(Dilated, Hypertrophic, Restrictive, Left Ventricular Non-Compaction or Arrhythmogenic Right Ventricular Cardiomyopathy)

Funding Opportunity:

The Children's Cardiomyopathy Foundation (CCF) is inviting investigator-initiated research applications for innovative basic, clinical, population or translational studies relevant to the cause, diagnosis or treatment of primary cardiomyopathy in children under the age of 18 years. Funding is available in the range of US\$25,000 to US\$50,000 for one year of total direct costs.

Eligibility Requirements:

The principal investigator must hold an M.D., Ph.D. or equivalent degree and reside in the United States or Canada. The investigator must have a faculty appointment at an accredited U.S. or Canadian institution and have the proven ability to pursue independent research as evidenced by original research in peer-reviewed journals.

Application Deadline:

Grant guidelines and application form are available online at childrenscardiomyopathy.org/site/grants.php. The 2011 deadline for application submission is September 2, 2011 with final award decisions to be made by January 2012.

For more information, contact
Lisa Yue, CCF president at
866-808-2873, ext 901 or
lyue@childrenscardiomyopathy.org

CCF Announces 2011 Research Grant Recipients *continued from page 3*

failure and sudden death but as a milder form in adults. This contrasting form of the disease between children and adults, also known as bimodal disease distribution, suggests that the primary DCM mechanism in childhood may change over time and that the normal changes associated with cardiac growth in children are potentially involved.

The Children's Cardiomyopathy Foundation has awarded \$150,000 in seed funding to three new research studies on pediatric cardiomyopathy for 2011.

Dr. Tardiff's research will focus on how changes in sarcomeric protein expression drives cardiac remodeling thereby altering the disease state. She believes that the bimodal and age-dependent features of the D230N mutation is due to changes that occur over time in a protein that interacts with Troponin T, an important Tropomyosin binding partner that contributes to the heart's contractile machinery. Dr. Tardiff expects that the way this mutant form of Tropomyosin (D230N) and Troponin T work together will be altered because of the mutation and lead to abnormal cardiac function. Furthermore, the disease is caused by an increased susceptibility of the cardiac muscle to injury in response to external stressors. "We need to understand what happens at the biophysical level – understand the function of the mutant gene product itself," says Dr. Tardiff. "Then potentially we can work to short-circuit the development of the disease."

Her research plan is two-fold – first to look at the molecular mechanisms of DCM and secondly to study the disease physiology. Initially, she and her team will build functional protein complexes and observe what changes occur when the mutant gene is a building block of

Tropomyosin versus when a normal form is included. She also will look at how Tropomyosin with and without the mutant gene interacts with Troponin T.

In the second part of her research, Dr. Tardiff will induce myocarditis, an infection of the heart, in three discrete groups of mice in order to understand the effect of mutant genes on the "stressed" heart. The first group of mice will have normal cardiac structure; the second group will have a mutant form of Troponin T, and the third group will have a mutant form of Tropomyosin. Dr. Tardiff expects only groups one and two of mice will survive the myocarditis "challenge" without any damage to the heart or with less damage than group three incurs.

"The ultimate impact of a study like this," says Dr. Tardiff, "is that it will tell us how to change the risk that children with cardiomyopathy, caused by mutant sarcomeric proteins, face in the long run. It will give us new ideas about appropriate interventions that will be effective in childhood."

STUDYING ARVD5 IN A HOMOGENEOUS POPULATION

Kathleen Hodgkinson, Ph.D.

assistant professor of clinical epidemiology and genetics at Memorial University of Newfoundland

Many young people with arrhythmogenic right ventricular cardiomyopathy are at risk of sudden cardiac death due to ventricular tacharrhythmias, heart failure and structural anomalies of both heart ventricles. Many families in Newfoundland, the Eastern most province in Canada, have a genetic subtype known as arrhythmogenic right ventricular cardiomyopathy type 5 (ARVD5), which can lead to sudden cardiac death (SCD) due to a mutation in the TMEM43 gene.

Although all adults with this mutation will show some sign of ARVD5 in their lifespan, it is not known who will develop

the lethal form of the disease, what the early effects are on children and whether the prognosis can be altered. Dr. Hodgkinson's study will look at a homogenous group in Canada to determine how the presence of the TMEM43 mutation clinically affects those under 18 years. This largely isolated "founder population," where there has been very little migration to and from the area, makes it ideal for genetic studies due to its ancestral purity and low genetic variation.

Dr. Hodgkinson plans to study a group of approximately 100 children from 18 large families between the ages of 10-18 who have one parent with ARVD5 to determine which individuals are at greater risk of developing the worst signs of the disease as they age. The study will involve following the children annually with routine cardiac clinical tests and adding two additional diagnostic tests using signal averaged ECG and tissue Doppler imaging of the heart.

The goal of the study is to understand what causes the mutation and the early-stage affects of ARVD5, which will provide important clinical information for diagnosis and prognosis. In particular, it can help guide doctors on when a child should be recommended for an implantable cardioverter defibrillator (ICD), the most common form of treatment for protection against sudden cardiac arrest.

"This ravaging disease does occur elsewhere in the world and what we learn here will inform cardiologists everywhere," says Dr. Hodgkinson. "This study and its results can potentially save the lives of children and their family members who are affected by ARVD5."



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Shelley Miyamoto, M.D.

Cardiomyopathy Specialist:

Shelley Miyamoto, M.D.

Shelley Miyamoto, M.D. is a pediatric cardiologist and director of the heart failure and cardiomyopathy program at the **Children's Hospital** in Aurora, Colo. Dr. Miyamoto first became interested in pediatric cardiomyopathy

when she was a fellow working with Mark Boucek, M.D., an expert in heart failure and a pioneer in infant heart transplantation. She cites his mentorship as an important factor in her decision to dedicate her career to caring for children with cardiomyopathy and researching the disease.

"One patient experience that continues to influence me is a 2-year-old girl who presented with severe heart failure," says Dr. Miyamoto. She was critically ill with dilated cardiomyopathy caused by viral myocarditis, and physicians were

not certain she would survive. However, she slowly improved and within one year her heart function had returned to normal and she was weaned off of all cardiac medications. "Witnessing the gratitude, joy and relief of this family at the recovery of their little girl impacted me greatly and has motivated me to continue with research that could lead to improved outcomes for children with heart muscle disease," continued Dr. Miyamoto

Dr. Miyamoto's research interest is on the cellular events leading to heart failure in children. She is a co-investigator on the study "Beta-adrenergic Receptor Regulation in Pediatric Heart Failure" and a primary investigator on "Cardiac Beta-adrenergic Adaptation in Pediatric Heart Failure." She believes that future research in cardiomyopathy and heart failure should focus directly on the pediatric population. "For the most part, the treatment of children with cardiomyopathy is extrapolated from what is known to be effective in adults with heart failure. But it is not clear that children respond to medication in the same ways as adults," Miyamoto noted.

Multi-Center HCM Study *continued from page 1*

President Lisa Yue. "We need to find this piece of the puzzle before we can consider clinical genetic testing and develop viable therapies. Families also want to have a better understanding of the risk of having another similarly affected child."

The most advanced genomic technology and genetic analysis methods will be used. Known as exome capture and sequencing, a portion of the DNA that contains our 20,000 genes is sequenced and analyzed. After sequencing the coding region of the genes, known normal genetic variants will be filtered out and disease-causing mutations in new genes for infantile cardiomyopathy will be identified. After identifying new genes for infantile cardiomyopathy, other children with cardiomyopathy will be screened for mutations in that same gene to confirm findings.

Once a set of genes for infantile cardiomyopathy is defined, the next phase of the study would be to characterize the diseases associated with these mutations, determine the frequency and types of mutations associated with cardiomyopathy, and compare the similarities and differences between infantile and non-infantile forms of cardiomyopathy.

"Identification of new genes for infantile cardiomyopathy should help establish new targets for treatment, clarify the prognosis for families, and provide reproductive options for families to have healthy children in the future," says Dr. Chung.

For more information or to enroll in the study, please contact Julia Wynn, genetic counselor at Columbia University Medical Center in New York City. She can be reached via phone, 212-305-6987 or email, jw2500@columbia.edu.

Recruiting Families for HCM Multi-Center Study

This multi-center HCM genetic study is currently recruiting patients in the U.S. that fit the below study criteria. The study will address commonly asked questions from families: why did this happen to my child, will my child have other problems beyond the heart, and will this happen again to someone else in my family or if we have more children?

Participation in the study requires a small blood sample from the child with cardiomyopathy and both parents. If there are multiple family members with cardiomyopathy, a blood sample will be requested from these affected family members as well. Blood can be obtained during a blood draw for routine labs at a doctor's office or a blood draw can be arranged at your home if you live in the U.S.

An explanation of the study will be given over the telephone and a brief family history will be taken. Copies of medical records also will be requested from your doctor to review your child's previously performed tests.

A family is eligible for the study if:

- Their child was diagnosed with hypertrophic cardiomyopathy at less than one year of age. The child may be older than a year of age now.
- Multiple family members are affected in that there is more than one affected child or both parent and child are diagnosed with cardiomyopathy. Such families are especially encouraged to participate.
- The specific cause of the cardiomyopathy has not been identified to be Noonan syndrome, a mutation in a sarcomeric gene, a metabolic disorder or Pompe disease.
- The family has already undergone genetic testing to rule out these causes and other systemic diseases that may cause cardiomyopathy.

FAMILY FUNDRAISERS

The Cinca Family Benefit for Cardiomyopathy: A Shopping Extravaganza

It was no secret Heather Cinca was having a fundraiser for CCF. She and friends let everyone know about their November 2010 holiday shopping event through fliers, advertising, letters and emails. The fundraising event became the talk of the town in Cocoa Beach, Fla. attracting 26 vendors and 150 attendees. The event raised more than \$12,000 for CCF.



The Cinca family & Brenda Grochowski speaking

Heather, a CCF family member since 2008 whose daughter, **Cristina**, was diagnosed with hypertrophic cardiomyopathy, had a strong desire to help CCF raise funds for research that could possibly improve treatment options for Cristina and other affected children like her. Heather and her friends **Brenda Grochowski** and **Dick Beagley**, came up with the idea of a yearly shopping event, “**The Cinca Family Benefit for Cardiomyopathy: A Shopping Extravaganza**,” in honor of Cristina.

The event included food and beverages, and attendees were treated to a song and dance performance from Cristina’s friends. Heather also invited local organizations for CPR demonstrations and to promote the importance of organ donation. Vendors, including **31 Gifts**, **Avon**, **Jockey** and **Creative Memory**, gave back a percentage of their sales, and other specialty booths enticed attendees with home made items such as treats baked by Cristina’s teacher and soda-can-tab jewelry made by Cristina’s Girl Scout troop. There was also a silent auction of donated gift baskets.

“One of the most important motivations for me is that this event makes Cristina feel like a rock star! She looks forward to it and gives a speech every year,” says Heather who has already scheduled this year’s event for November 20.

Heather’s Tips for Fundraising Success

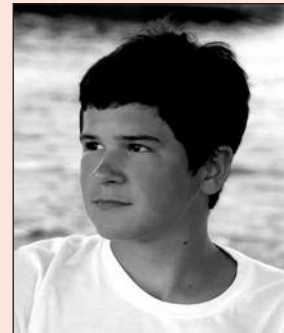


Heather Cinca & Brenda Grochowski

1. **Start planning early.** An event like this requires at least two months of planning.
2. **Plan the event with a group** of people who can provide good advice and assistance.
3. **Advertise** in as many places as possible and as far in advance as possible.
4. **Spread the word early** through solicitation letters, invitations, conversation, and email.
5. If you are having a shopping event, **bring in many vendors** and offer as wide a variety of products as possible.

More Family Fundraisers...

Tee Off for Kyle



Kyle Rymiszewski

Ken Rymiszewski held a golf outing among friends and colleagues to honor his son, **Kyle**, who lost his battle to hypertrophic cardiomyopathy in December of 2009. Kyle was a sports enthusiast

and one of his proudest moments was being awarded Most Valuable Player while playing for the Michigan All Star Baseball Team. To date, the Rymiszewski and Cowher families have raised more than \$30,000 for CCF through memorial gifts and fundraisers for Kyle. Another golf outing is planned for July 16.

Hope For Little Hearts Dinner Auction

For the fourth year running, **Melissa Sabin’s Hope For Little Hearts Foundation** organized another successful fundraiser for CCF, raising more than \$7,700. The event was planned in honor of her son **Brody**, age 6, who has left ventricular non-compaction cardiomyopathy, and featured a BBQ dinner and auctions at the Swiss Park in Bonney Lake, Wash. An enthusiastic crowd of friends and family took part in both the silent and live auctions, which featured an array of donated products and services from the community. Melissa has a fun run coming up in June and has already started planning for next year’s dinner event!



The Sabin family

Donating to CCF just got better!

Amanda DiMonda of **DiMonda Photography**, was diagnosed with peripartum cardiomyopathy after her second child. Each year she donates 10 percent of her holiday print sales to CCF. This year she is taking it one step further and giving a \$50 print credit on a photography session to anyone who donates \$50 or more to CCF. For more information, contact Amanda at dimondaphotography.com or call CCF at 866-808-2873 ext. 903.

Honor Students Supporting CCF

The **National Junior Honor Society** of **Loggers' Run Community Middle School** in Boca Raton, Fla. chose CCF as one of ten nonprofits to support with proceeds from their holiday candy cane and Valentine's Hershey's Kiss sales. CCF was selected by one of the student members whose friend has pediatric cardiomyopathy.



Melisa Salamony-Fulling (center) and colleagues

her husband **Curtis** and two sons **Cameron** and **Collin** who have hypertrophic cardiomyopathy. Melisa donated \$1 to CCF for every colleague who wore red, and her OnSight supervisors generously matched her donation.

A Holiday Concert and Hoops for Heart Month

Sharon Tramm honored her daughter **Kristina**, who passed away from dilated cardiomyopathy in 2001, with two fundraisers: a 2010 holiday concert, "**Honoring the Heart of an Angel**" at King of Kings Lutheran Church in New Windsor, N.Y., and a February school event at the Balmville Elementary School in Newburgh, N.Y. Students raised funds through coin collections and a "**Hoops for the Heart**" basketball free throwing contest. Sharon also arranged a school assembly to educate students on cardiomyopathy, CCF's mission and the importance of physical activity and a healthy diet.

Will CCF Be Mine

The **Tyngsborough Elementary School** in Massachusetts held a **February Heart Month** fundraiser in honor of Assistant Principal **Scott Middlemiss'** son **Joe** who has hypertrophic cardiomyopathy. Led by teachers Erica Yandow and Jen Trischitta, the school's student council made and sold valentines to students and staff for Valentine's Day.

Wearing Your Heart on Your Sleeve

Melisa Salamony-Fulling challenged her colleagues at **OnSight Health Care**, which has locations in Ohio, Michigan and Pennsylvania, to wear red for a day in support of



Mary Mauro

Volunteer Spotlight: MARY MAURO

Finding Time In Your Heart

Mary Mauro, executive director at **Morgan Stanley**, remembers fondly when her father, a veteran NYC police officer, would ask her and her two

brothers to write essays on how they helped someone in need. Her father's emphasis on community service left an indelible mark on Mary, and at one point in her career she volunteered at nine charities. She learned about CCF through Eddie Yu, CCF founding board member, while working at Morgan Stanley. She gladly offered to help at CCF's fundraisers. "There are other organizations that are bigger and with more recognition power, but CCF's cause is just as important and worth championing," said Mary.

Mary's bubbly personality is now a mainstay at CCF's poker and golf events. She is the consummate hostess and makes all who attend feel welcome. CCF is lucky to have her as a volunteer. "It feels great to give back and volunteer for CCF," said Mary. "In some small, small way, I hope I am making a difference."

CCF is Four-Time Beneficiary of Medtronic Global Heroes Award

The **Medtronic Foundation** recently awarded \$1,000 to CCF on behalf of its 2010 Global Hero, **Pablo Maillet** of Santiago, Chile. Every year, the Medtronic Foundation celebrates the passion and accomplishment of runners whose lives have been improved by medical devices. 25 such athletes from around the world are invited to participate in the Medtronic Twin Cities Marathon in Minnesota. For the fourth consecutive year, CCF was selected as the beneficiary patient organization by a Medtronic sponsored runner.



Pablo Maillet, Medtronic Global Hero

Pablo Maillet was diagnosed with a congenital atrioventricular block at the age of two and became one of the youngest patients in Chile to receive a pacemaker. The device allowed him to play soccer and trek through the Andes Mountains as a young boy, and eventually take up running to be part of the Medtronic TC 10 Mile event.

CCF POKER EVENT Deals a Winning Hand Third Year in a Row



Board member Eddie Yu and tournament winner Kevin Tan

On Wednesday February 9, CCF hosted its **Third Annual No Limit Texas Hold 'Em Poker tournament** at **Crimson** in New York City. With nearly 250 attending the sold-out event, more than \$213,000 was raised for CCF's research and education initiatives.

"The tournament just keeps getting better, and this year we raised 70 percent more from the event than in our first year," said CCF Board member **Carney Hawks**. "We are truly grateful for the support of our donors, who are helping us to continue our mission of finding a cure for pediatric cardiomyopathy."

Guests and players from top law firms and financial companies in the tri-state area enjoyed cocktails and a light supper

in the early evening followed by a rousing game of poker that lasted well into the night.

The night's grand-prize winner **Kevin Tan** of the **Macquarie Group** won a \$10,000 entry to the 2011 World Series of Poker Main Event in Las Vegas. Kevin is looking forward to trying his luck at the famed tournament in July. "The CCF poker event was great fun for a great cause. I will definitely look to participate again next year," said Kevin.

Second prize of an all-inclusive, three-night trip for two to Las Vegas went to **Jordan Freundlich**. **Ken Senior** won the third prize, an Atlantic City getaway at the Borgata Hotel Casino and Spa. Fourth prize of golf and lunch at Hudson National Golf Club went to **Michael Schwartz**. **David Key** was the fifth-prize winner, four season tickets to a New York Giants game. **James Higgins** and **Carney Hawks** won sixth and seventh prizes, dinner gift certificates to Union Square Café. Eighth prize, a gift certificate to Hearth Restaurant in New York City, went to **Greg Steele**. **Bryan Wolff** and **Jed Walsh** won the ninth and tenth prizes, gift certificates to Tribeca Wine Merchants.



Top photo: Playing poker at Crimson

Bottom photo: Matt Carter, Brian Lee, Peter Taukus, Dan Ornstein



SAVE THE DATE!

Montclair Golf Club
West Orange, New Jersey

Monday July 18, 2011

Join CCF for this popular and fun-filled event!

Visit our event web page under "News & Events" for more information.

2011 Event Sponsors: BGC/Mountain; Brigade Capital Management; BTIG, LLC; Benji Cheung/Bob Franz/Jed Kelly; Dechert, LLP; Chris Delong; David Fisch/Gladys Golden/Barbara Mansdorf; John Florio/Andy Stock; Eric Friel; Rob Gallivan; Gibson Dunn & Crutcher; Bill Goebelbecker; Goldman Sachs; ICAP High Yield; Imperial Capital; JP Morgan High Yield/ Gene Pagnozzi; Katten, Muchin & Rosenman; Norm Louie; John Lugano/ Peter Schellbach; Matt Philo; RBS; Darren Richman; Riva Ridge Capital Management; Steve Rosen; The Sandler Family; Tullett/Chris Berry/Matt Lambert; Vanderbilt Appraisals; The Yu Family

More event photos are online under "News & Events/Poker Event."

Genetic Diseases of Children Conference Held in New York City



Panelists Wendy Chung, Lisa Yue, Gina Cioffi, David Lynch and Marshall Summar.

Lisa Yue, CCF president and founder, and **Kella Boyer**, patient outreach and support manager, recently attended the “**Genetic Diseases of Children: Advancing Research and Care Conference**,” hosted by the New York State Department of Health.

Held in New York City in March, this national conference brought together a diverse group of researchers and clinicians, patients and caregivers, patient organization advocates, and government

and industry leaders to focus on the unique health care challenges faced by children living with a genetic disease. State Health Commissioner **Nirav R. Shah, M.D., M.P.H.**, said, “The goal of this conference is to develop recommendations for establishing a comprehensive, state-based model that will effectively facilitate early diagnosis through newborn screening, improve access and quality of care, optimize coordination of provider services, and foster high-impact clinical research to expedite improved medical treatments.”

The conference was organized into four plenary sessions with over 35 panel discussions covering early diagnosis, family experiences, care and coordination of services, meeting the needs of children with a genetic disorder, and accelerating research. Lisa was invited to be a panelist on “Optimizing Care: Lessons Learned” moderated by **Wendy Chung, M.D., Ph.D.** Other panelists included **Gina Cioffi, Cooleys Anemia Foundation**; **David Lynch, M.D., Ph.D., Children’s Hospital of Philadelphia**; and **Marshall Summar, M.D., Children’s National Medical Center**. This section highlighted the practical methods for advancing care of children with rare conditions when there is variability in evaluation and treatment, and little scientific evidence to guide medical management. Topics discussed ranged from assisting families in locating a specialist and center of excellence to educating physicians on treating genetic diseases in a standardized and more comprehensive manner.

ICD and Sports Participation Study Underway

A nationwide study lead by **Rachel Lampert, M.D.** of **Yale University School of Medicine** and in collaboration with other heart specialists is being conducted on the safety and risk of sports participation for individuals with implantable cardioverter defibrillators (ICDs). The safety of sports participation for individuals with ICDs is largely unknown. Because it has not been reported how well an ICD will work during sports and the possibility of injury, most physicians advise patients with ICDs to refrain from participating in any sports more vigorous than golf.

A recent survey of physicians caring for patients with ICDs suggests that, despite the current physician guidelines, some patients with ICDs still participate in sports. The goal of this multi-center study, known as the ICD Sports

Safety Registry, is to determine whether ICDs work as effectively under the stress of competitive sports. The study will help answer questions such as do athlete’s hearts need shocking more often when participating in intense sports, would the ICD fire hard enough to overcome the physical duress and surge of adrenalin, and could a sport’s repetitive motions or a hit to the chest break the ICD.

The study is currently recruiting people with ICDs age 10-60 who are participating in competitive sports and then following them prospectively for four years. To date, more than 300 athletes have been enrolled.

For more information or to enroll in the study, please visit icdsports.org or call the registry coordinating center at 866-207-9813.

Q&A with Jenny Nova, RN, CCTC:

Teens and Medication Compliance



Jenny Nova is the pediatric heart failure & transplant practice nurse and clinical transplant coordinator at Montefiore Children's Hospital in the Bronx, New York. She has worked with hundreds of children with complex heart disease and heart failure and coordinated care for more than 200 children undergoing heart transplant. Based on her 12 years of experience, Jenny shares her insights on teens and medication compliance.

Q: How often is medication compliance an issue for teenagers who have cardiomyopathy or have received a transplant?

A: Medication non-compliance in the adolescent population is a challenge. It is difficult to quantify the prevalence since patients do not always disclose it. Patients who have received a transplant require frequent monitoring of their immunosuppressive drug levels, and sub-therapeutic levels can be an early clue that compliance may be an issue.

Q: Why do teens sometimes neglect to take their medications?

A: The dynamics and reasons for non-compliance in the adolescent are poorly understood. This stage of development is influenced by their feelings of invincibility. This clouds their ability to connect the consequences of their actions when they skip their medications. We know that risk factors include lack of family support, low level of understanding about their disease and denial. They also may simply forget. Also, teenagers are influenced by their peers. For instance, they might feel "different" if they have to break away from their friends to take medications, or they may dislike having to use the bathroom often due to their diuretic.

Q: What are the consequences of missing medication doses?

A: It depends on the medication. Immunosuppressant medications are prescribed after a transplant and are critical in preventing the body from rejecting the new heart. If they are not taken daily and on a regular schedule the patient can experience rejection.

In patients with cardiomyopathy or heart failure as a result of cardiomyopathy, the goal of medication is to decrease the workload of the heart. They also may be on diuretics to help them eliminate extra fluid in their lungs. If a patient stops taking these medications they may develop congestive heart failure.

Q: What can professionals do to encourage teens to take their prescribed medications?

A: Developing a good relationship is vital in getting patients to at least "listen" to you. You must include them in the discussions about their disease and treatment and make them feel as part of the medical team. It is important for them to trust their medical team and know that we are here to support them no matter what. I often counsel my adolescent patients on strategies to improve compliance, such as having them develop a medication schedule that fits their lifestyle.

We often recommend the use of cell phone alarms to remind them. In some

cases, we reach out to the school nurse and ask if they can administer the morning doses of medications in school.

Q: What can parents do to help ensure their teens take their medications regularly and correctly?

A: Just like physicians and the rest of the medical team, parents must establish an open relationship with their child. It is a very frustrating process but keeping the lines of communication open is vital in getting them through the "rough patch." They should also use modern technology to their advantage. Instead of telling them to take their medications, send them text reminders on their cell or via email. If timing of the medications does not work for them, explore options with your medical team. Sometimes a simple adjustment can make a positive impact, as all the teen wants is some sense of control over their lives.

Q: What can be done to help with medication compliance as an adolescent transitions into adulthood?

A: Educating the adolescent patient about their disease process is fundamental. It helps to establish a foundation for their transition to self-care. As they get older and more independent, most teens should be encouraged to take charge of their own care. Parents are advised to transition them slowly and gradually, as the teen needs to prove that they can do it on their own. You can start with something as simple as having them call to make their next doctor's appointment.

During the transition, it is important for parents to supervise their teen without crowding or overwhelming them. This process is gradual and becomes increasingly important when teens are planning to go away to college. During the year prior to college, the parents and medical team need to counsel the teen about all aspects of college life that will affect their compliance such as irregular sleep habits, increased peer pressure and the physical demands of college life.

A Teenager's Perspective: *Living Life On Your Own Terms with HCM*

Ben Johnson definitely knows something about living with a chronic heart disease. During a routine 3-month-old well visit, Ben was diagnosed with hypertrophic cardiomyopathy (HCM). At the age of



Ben Johnson

11, he had a myectomy to remove part of his septum and three years later, after nearly fainting while walking up a flight of stairs, he had a defibrillator implanted at the age of 14. Today, he is a freshman at Michigan State University and facing his diagnosis head on, and managing his life with HCM on his own terms.

"Because I was diagnosed so young, I didn't really feel restricted by the disease," says Ben who participated in all school activities except gym class. "My parents specifically scheduled activities so that I wouldn't strain myself, but they never coddled me." Ben is thankful his parents, Matt and Rhea Johnson, gave him the space he needed to test and find his physical limits.

When Ben had his defibrillator implanted as a teen, he felt challenged by the disease for the first time. "This affected me even more than open-heart surgery even though it is a far less intense procedure," admits Ben. "I felt like it would make me stick out even

more, and every teenager wants to fit in."

For Ben the ease with which he accepted his diagnosis came as he began to realize he would have to live with the disease and the restrictions that came with it for the rest of his life. Naturally, as a teen, his focus was

on how the disease would impact his friendships. He admits he felt nervous about telling his friends, but fortunately he found support with many. "I found that because I couldn't relate with people on a physical, athletic level, I became friends with people who I could relate to on a more personal level," he says. Instead of sports, Ben found more creative outlets such as Men's Choir, Jazz Band and Quiz Bowl.

Throughout high school Ben's parents offered encouragement, support and the space to become his own person, which provided him with the confidence to take charge of his HCM. Once he began driving, he would stop by the pharmacy on his way back from school to pick up his prescriptions. At doctor's appointments, his parents had him fill out his own forms, which gradually led to scheduling and attending his own appointments. Now that he is away from home, doing such things independently feels natural.

One challenge for Ben and many teens

with cardiomyopathy, is to adhere to a strict medication schedule, but Ben says with practice it has become as automatic as getting out of bed in the morning. "When you're just starting to take your medication, keep track of it in a very obvious way," he says. "I have heard of many techniques for remembering. A girl who enjoyed knitting made a small purse to keep her meds in. Another guy I know programmed an alert into his computer; a former athlete thought of it as his new work-out schedule."

"My biggest tip for teens and parents is openness," says Ben. "For example, a parent might be hesitant or even scared to let their teen manage their medication, but it is an important step to a mature and trusting relationship. By giving me a voice in my healthcare, my parents have taught me to be my own best advocate."

His motto in managing HCM and life is to focus on what you can do and not on what you cannot do. "I can't run a 4x4 on the track team, but I can play a mean guitar," says Ben. "The only things holding you back are the limits of your desire." Ben is majoring in anthropology in the hope of one day working in medical anthropology to improve medical systems and the culture of medicine. His story is proof that, with confidence in yourself and the support of loved ones, it is possible for teens to live with cardiomyopathy on their own terms.



Miriam Kaufman, M.D.

BOOK REVIEW:

EASY FOR YOU TO SAY: Q&As for Teens Living with Chronic Illness or Disability

The teenage years can be difficult, and when you are a teen with a chronic illness normal life issues are magnified and become much more challenging. In the book, *Easy for You to Say: Q&As for*

Teens Living with Chronic Illness or Disability, **Miriam Kaufman, M.D.**, medical director of the complex adolescent problem program at **Toronto Hospital for Sick Children**, offers sound advice to questions posed from teens with chronic illness.

Dr. Kaufman, who has spent three decades working with teens, has a "strong belief in fighting for what you need and want," and throughout the book her responses teach self-empowerment through open and honest communication.

Originally published in 1995, the book was updated in 2005 to cover additional topics and concerns. All teen questions are from Dr. Kaufman's work encounters and research. The book, which is in an easy-to-read question and answer format, is divided into eight sections: Family Relationships; Doctors and Medical Illness; Friends and Dating; School and Work; Alcohol, Drugs and Medication; Sexuality; Recreation; Transition. She also tackles difficult subjects such as birth control, recreational drug use and death. The appendix includes a glossary of useful terminology for teens, details information about drug interactions and side effects, and has a resource section listing websites on teen health, travel and sexuality.

Easy for You to Say: Q&As for Teens Living with Chronic Illness or Disability is relatable to any teen with a chronic illness and serves as a guide for those who work or live with teens seeking self awareness and acceptance of their condition. As Dr. Kaufman emphasizes, the desire for acceptance, independence, and for a better understanding of and control over one's health, are universal desires.

HearttoHeart

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The Newsletter of the Children's
Cardiomyopathy Foundation

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MEMBER SUPPORT SERVICES

CCF offers a variety of ways for members to learn more about pediatric cardiomyopathy, share information and provide support to one another.

For more information on the below scheduled events, contact Kella Boyer at kboyer@childrenscardiomyopathy.org

CCF Forum Guest Q&A Sessions

- **Maintaining a Committed Relationship**

*Melissa Groman, L.C.S.W.,
Therapy Associates, LLC
May 31 – June 6*

- **Automated External Defibrillators**

*Rachel Moyer, Founder
Greg W. Moyer Defibrillator Fund
August 15 – 22*

- **Cardiomyopathy Genetics**

*Wendy Chung, M.D., Ph.D.,
Director of Clinical Genetics
Columbia University Medical Center
September 12 – 19*

Local Support Groups

- **Florida**

Jackson Memorial Hospital, Miami, Fla.
May 21 at 1:00 p.m.
August 27 at 1:00 p.m.
October 22 at 1:00 p.m.
Diagnostic Treatment Center, Room 270

- **Illinois**

**Loyola University Medical Center
Maywood, Ill.**
May 14 at 11:00 a.m.
Stritch School of Medicine, Room 170

- **North Carolina**

**Duke University Medical Center
Durham, N.C.**
April 28 at 6:30 p.m.
May 26 at 6:30 p.m.
Room 4902, McGovern-Davidson
Children's Health Center

- **Ohio**

**Cincinnati Children's Hospital
Cincinnati, Oh.**
May 18 at 6:00 p.m.
Room EDA 603
Liberty Township Campus
September 24 at 11:00 a.m.
Cincinnati Children's Hospital
Medical Center

Phone Support Group

- **General Discussion**

May 26 at 8:00 – 9:00 p.m. Eastern
June 23 at 8:00 – 9:00 p.m. Eastern
July 21 at 8:00 – 9:00 p.m. Eastern
August 25 at 8:00 – 9:00 p.m. Eastern
September 22 at 8:00 – 9:00 p.m. Eastern

- **Panel Discussion with Young Adults**

Featuring a panel of several young adults living with cardiomyopathy or with a heart transplant.
May 18 at 6:30 – 7:30 p.m. Eastern