First CCF-AMERICAN HEART ASSOCIATION Grant Recipient Announced

In 2007, the Children’s Cardiomyopathy Foundation and the American Heart Association (AHA) jointly established the Pediatric Cardiomyopathy Research Award. This July, CCF and AHA selected the first grant recipient, Dr. George Porter, Assistant Professor of Pediatrics, Pharmacology and Physiology at the University of Rochester. He will be awarded $198,000 over a two year period by CCF and AHA jointly.

In his research to date, Dr. Porter and his team have discovered that the deletion of CaV1.2, the major calcium channel in the heart, causes non-compaction cardiomyopathy in the embryo. Non-compaction cardiomyopathy is a rare form of cardio-myopathy that occurs when the muscular wall of the left ventricle is not properly compacted and there are abundant muscle protrusions and deep recesses (trabeculations) in the ventricular wall.

In his CCF-AHA funded study, Calcium Channel Disruption, A New Model of Non-Compaction Cardiomyopathy, Dr. Porter suggests that abnormal calcium

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CCF PARTNERS with the PEDIATRIC CARDIOMYOPATHY REGISTRY to FUND TWO STUDIES

This year, CCF will work with the National Institutes of Health sponsored Pediatric Cardiomyopathy Registry (PCMR) to fund two studies utilizing patient data from the registry. The studies will focus on two areas - growth patterns and serial echocardiographic measures - and examine the relationship of each to disease severity and outcomes in children with cardiomyopathy.

The first study, The Relationship of Growth Patterns on Outcomes for Children with Cardiomyopathy, conducted by Tracie L. Miller, MD;

Sarah E. Messiah, PhD; Steven E. Lipshultz, MD; and E. John Orav, PhD will examine the effects of growth failure, one of the most significant clinical problems of children with cardio-myopathy. Growth failure can occur when severe cardiac dysfunction results in poor absorption of food and increased metabolism. This malnutrition can aggravate the cardio-myopathy. Thus

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A Cause for Today...A Cure for Tomorrow
From Lisa Yue,
CCF Founder & President

Continuity gives us roots; change gives us branches, letting us stretch and grow and reach new heights.
— Pauline R. Kezer

Fall is a time for changes – a new season, a new school year. Now is also a time of positive change for CCF. After serving as the Executive Director of CCF for six years, it is my pleasure to announce that Saul Spivack, PhD has joined us as CCF’s new Executive Director. I will be moving away from the Foundation’s daily operations and redirecting my efforts to overseeing the Board of Directors, working on special projects and cultivating strategic alliances in support of CCF’s vision.

Since our establishment in 2002 we have grown to an organization with a $700,000 budget and $1.3 million in assets, while reaching out to 800 families worldwide and committing over $850,000 to research and treatment initiatives. As we prepare for our next phase of growth, Saul’s experience will help CCF move more formally into that phase.

Saul’s career has focused on improving healthcare delivery and outcomes in corporate, philanthropic, government and academic settings. He has worked at companies and institutions as diverse as the University of Pennsylvania School of Medicine, Metropolitan Life Insurance, Towers Perrin Consulting, Price Waterhouse and Pfizer. Saul was most recently the Executive Director of Sage Eldercare, a nonprofit provider of long-term health, social support and educational services to seniors and their caregivers.

As we move forward with our changes, we look forward to your input and your continued support.

Lisa Yue

First CCF - AHA Grant Recipient Announced  continued from page 1

channel function causes abnormal ventricular myocardial organization, leading to non-compaction. The goals of this study are to better understand what is being disrupted in the heart to cause this form of cardiomyopathy and then to determine the effects of non-compaction in young patients. Dr. Porter believes that “the results may help to figure out ways to prevent and treat non-compaction once it is detected.” They could also help guide and encourage genetic testing, risk assessment, reproductive counseling and treatment for patients with this form of cardiomyopathy.

To test his idea, he will use four mouse models in which he will delete CaV1.2 from cardiac myocytes at different stages of their development in the mice. He will then use qualitative methods to study changes in myocardial structure, cardiac function and cardiac rhythm in each model. Finally, he will perform experiments to determine how intracellular calcium signaling pathways may regulate compaction.

Dr. Porter believes that “the results may help to figure out ways to prevent and treat non-compaction once it is detected.”
In April 2008, the New England Journal of Medicine published a study, *Shared Genetic Causes of Cardiac Hypertrophy in Children and Adult*, that clarifies some of the genetic assumptions of hypertrophic cardiomyopathy (HCM) in children. For years, scientists assumed that adult HCM was clinically distinct from pediatric HCM. In a study led by Drs. Jonathan and Christine Seidman of Harvard Medical School, it was found that adults and children with HCM have several gene mutations in common. This discovery will impact the development of future diagnostic tests for pediatric HCM.

The Seidmans collaborated with pediatric cardiologists Dr. Amy Roberts of Children’s Hospital Boston and CCF funded investigator Dr. Jeff Towbin of Baylor Medical College to gain insight and access to young HCM patients. Dr. Towbin was awarded funding by CCF in 2006 to conduct several genetic studies on pediatric cardiomyopathy. Some of his findings were utilized in this collaborative study.

The scientists extracted DNA from children with idiopathic HCM. The team studied the ten genes that are responsible for adult HCM and discovered that some children harbored the same mutations in them. Only 33 of the 84 children studied had a family history of HCM. In 64% of those children, mutations were identified. Of the majority that did not have a family history, mutations were still identified in nearly 50% of the children.

The team then took a closer look at the genes of parents of the affected children without a family history of the disease. In seven of the 11 cases, one parent exhibited the same mutation as his or her child. Echocardiograms later revealed that the parent had HCM. Further genetic testing of this subgroup of children and their parents could reveal why some children present symptoms of HCM so much earlier than their affected parents, and this information could help to determine appropriate treatment. Says Christine Seidman, “This study demonstrates that kids who present with sporadic cardiac hypertrophy deserve the same genetic test as adults.”

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**CCF GRANT RECIPIENT Presents at AMERICAN HEART ASSOCIATION Scientific Session**

CCF’s 2007 grant recipient, Dr. T. Y. Hsia of University of South Carolina will present his preliminary research findings at the 2008 American Heart Association (AHA) Scientific Session in New Orleans on November 7th.

His findings are important in that they may explain why the clinical course and prognosis for children with DCM is so much worse than in adults. Dr. Hsia is the sixth CCF funded researcher to present at the premier cardiovascular forum.

Dr. Hsia’s abstract, *Differential Expression in the Determinants of Extracellular Matrix Remodeling in Pediatric and Adult Dilated Cardiomyopathy*, will show that there are distinct differences in the regulation of the heart’s extracellular matrix in children with dilated cardiomyopathy compared to adults. The extracellular matrix provides the scaffolding upon which the cells of the heart build, and its maintenance is critical for the heart’s proper structure and function. The extracellular matrix is regulated and maintained by two enzymes, the matrix metalloproteinases (MMPs) and their inhibitors (TIMPs).

In his study, Dr. Hsia discovered that in pediatric DCM patients there was a significant increase in a specific type of MMP, MMP-8, which can degrade all healthy components of the extracellular matrix. There was also a significant decrease in its inhibitor, TIMP-1. This exaggerated MMP-8/TIMP-1 ratio is unique to young DCM patients. It radically changes and destabilizes the extracellular matrix, causing the heart to eventually become thinned and not function properly.

The resulting manuscript of Dr. Hsia’s research will be published in the AHA journal, *Circulation.*
NEW RESEARCH into a Drug That Might HELP DIMINISH THE DEVELOPMENT OF HCM

Dr. Carolyn Ho, Medical Director of the Hypertrophic Cardiomyopathy Clinic at Brigham’s Women’s Hospital, is partnering with Dr. Steve Colan of Children’s Hospital Boston to conduct a pilot study on whether the drug Diltiazem, a calcium channel blocker, can decrease the progression of hypertrophic cardiomyopathy in adults and children. A recent study suggests that asymptomatic individuals who carry the gene mutation which causes HCM in their family may be able to take medications early in life to try to decrease changes to the heart caused by the mutation and prevent HCM from developing. Dr. Ho’s research will elaborate on these findings.

Dr. Ho’s study was initiated after promising results from HCM studies on mouse models performed at Brigham and Women’s Hospital. The mice were engineered to have the same type of gene mutations that cause HCM to develop in people later in life, typically as juveniles or young adults. In the study, it was found that mice treated with the calcium channel blocker, Diltiazem, early in life before they developed thickening of the heart muscle or other symptoms of HCM, had less heart muscle thickening and less microscopic changes compared to mice given a placebo.

Dr. Ho is looking to enroll adults and children (age 6 and up) who have a known sarcomere gene defect associated with HCM but have no symptoms and no thickening of the heart muscle on echocardiography in her study. For more information on enrollment, please contact Libby Sparks, RN at 617-432-1006 or lsparks@genetics.med.harvard.edu.

A recent study suggests that asymptomatic individuals who carry the gene mutation which causes HCM in their family may be able to take medications early in life to try to decrease changes to the heart caused by the mutation and prevent HCM from developing.

CCF Partners with the Pediatric Cardiomyopathy Registry continued from page 1

cardiomyopathy may lead to growth problems, but growth problems can lead to further complications that may directly or indirectly impact heart function.

The study will describe the growth patterns among the PCMR patients and determine how closely related abnormal growth among these children is to the form of cardiomyopathy that they have. It will also examine how variables such as rates of hospitalization, mortality, time to transplant and specific echocardiographic measurements impact growth failure. This information can help physicians identify patients who would benefit from transplant or other types of medical treatment and focus attention on early nutritional interventions that may prevent or delay further decline in heart function.

The second study, Serial Echocardiography as a Research Tool for Pediatric Cardiomyopathy Outcomes, is being conducted by Sarah E. Messiah, PhD; Steven E. Lipshultz, MD; E. John Orav, PhD; James D. Wilkinson, MD, MPH; and Jorge A. Alvarez. The study will investigate how changes in a series of echocardiographic measures might predict poor outcome (death and transplant) in infants and children with different forms of cardiomyopathy.

Existing PCMR echocardiographic data of different forms of cardiomyopathy will be analyzed to determine the extent, prevalence and progression of heart dysfunction and then compared to the clinical and demographic characteristics of children measured at the time of diagnosis. These findings will help identify those groups of children who are most likely to have persistent or worsening cardiac function based on identified echocardiographic patterns and therefore assist physicians in determining their medical management.

The Pediatric Cardiomyopathy Registry (PCMR) was established in October 1995 to collect data on pediatric cardiomyopathy patients 18 years of age or younger. The PCMR includes baseline and longitudinal data of over 3,000 children with primary cardiomyopathy collected from 98 hospitals in the US and Canada.
Sixth Annual **Golf Classic Raises Nearly $400,000 for RESEARCH**

This year’s Golf Classic took place for the second time at the historic and beautiful Montclair Golf Club in West Orange, NJ on July 21st. This year’s event was again a sold out event. 176 attendees from top name law firms, hedge funds, asset management firms, and other financial service companies enjoyed a day of great weather, challenging golf and good food. Despite the economic strains felt by all this year, the event still raised $394,911. The event proceeds will be earmarked for CCF’s research grant program and scientific conference.

At dinner, prizes were given to the winning foursomes which included:

- **Rich Brennan, Courtney Carson, Bill O’Donnell, Chris Reddy of Camulous Capital** (First prize);
- **Joe Mannello, Rich Rossi, Norm Kopack, Brain Taddeo of Broadpoint** (Second prize);
- **Chris Biel,**


Brian Reid, Pat Lannigan, John Heffers of KBC Financial (Third prize); and Brain Charters, Gene Pagnozzi, James Fitzpatrick, Laurent Renard of JP Morgan (Fourth prize). Prizes for the Closest to the Pin went to Joe Satto and Bob Frahm. The Longest Drive prizes went to Gerard Hickey and Seth Bernstein.

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**New Funding Sources Needed**

CCF is searching for new contacts for corporate donations and foundation grants. Last year, CCF raised over $94,000 from corporate donations, matching gifts and corporate foundation grants – many of which were made possible by CCF donor referrals and nominations.

If your company has a corporate giving program or a foundation that provides program grants, please let us know. Contacts to pharmaceutical companies, medical device companies or other charitable giving programs are also very helpful.

Please contact Pauline Pierrot, Assistant Director, at ppierrot@childrenscardiomyopathy.org with any personal contacts and/or referrals you may have.
CCF UPDATES

Second CCF CUT-A-THON Takes Place in California

CCF parent Fran Cole, hosted a Cut-A-Thon with Vibes Salon, on September 21st in Brentwood, CA in honor of her two sons Ben, 12 and Josh, 10. Over 200 people attended to help raise more than $2,000 for CCF’s research fund. Last year, Ben and Josh were diagnosed with hypertrophic cardiomyopathy and implanted with defibrillators. “Although it was difficult for us as parents to find out our two sons have HCM we want to be proactive and do whatever we can to help find a cure.”

Outside the salon, kids enjoyed getting kids tattoos and cookies, a Nintendo Wii raffle and a lemonade stand run by Ben and Josh. Cold Stone Creamery and Willy’s Bagels & Blends participated and donated 20% of their sales to the fundraiser. Fran hopes to host another CCF fundraiser this winter. “We want to keep doing what we can to raise money for research and help our boys and all other children with cardiomyopathy,” Fran adds.

SALEM ROAD RACE

For the third year in a row, Rick Konon organized the Salem Road Race to increase public awareness of pediatric cardiomyopathy and raise funds for CCF. An avid runner, Rick plans the event every year as a tribute to his nephew, Aidan, who has hypertrophic cardiomyopathy. On April 12th, 220 runners participated in the 5K community run in Salem, CT which was well supported by local businesses.

And More Family Fundraisers...

• In March, CCF family member Denise Gilroy nominated CCF for a fundraiser at Clarkstown High School South in West Nyack, NY. Once a month, the staff has an event called “Clarkstown Goes Casual for a Cause” which is a dress down day for the faculty and staff members. Contributions made on this day went to CCF.

• Melissa Witt and Sharon Belles hosted a Silent Quilt Auction and Sweet Heart Ball fundraiser in honor of Melissa’s 3-year-old daughter who has DCM. The February 23rd event was held at the Quilted Cottage quilt shop in Saginaw, MI and raised $2,000 for CCF.

Of Interest...

Call-to-Action to Improve Sudden Cardiac Arrest Survival Rates

Sudden Cardiac Arrest (SCA), commonly associated with certain forms of cardiomyopathy, is one of the leading causes of death in the United States. According to the American Heart Association, there are between 1,900 and 14,000 out-of-hospital sudden cardiac arrests among children annually. On average, less than 7% of those children survive.

In April, the National Medical Association (NMA), an organization focused on eliminating disparities in healthcare, held a meeting on SCA to highlight this under-recognized public health problem. Attended by the nation’s leading cardiac care professionals, government officials and patient advocates, the conference developed six directives to address the issue. The directives recommended increasing awareness of SCA among the community and healthcare professionals and encouraging policy makers to develop a plan of action that will establish SCA as a public health priority. The directives also called for developing SCA prevention measures, decreasing health disparities associated with SCA among ethnic groups and increasing support from insurers on SCA diagnosis, prevention and treatment.

To read the conference’s white paper, “Sudden Cardiac Arrest: Advancing Awareness and Bridging Gaps to Improve Survival,” visit www.nmanet.org/index.php/President/presidents_office/.
Screening Pilot Program to Kick Off in Texas Schools

Many children with cardiomyopathy do not show symptoms and remain undiagnosed while others are only diagnosed after an unfortunate incident like sudden cardiac arrest (SCA). The Early Cardiovascular Detection Pilot Program, formed by the Texas Education Agency (TEA) and created by Senate Bill 7, will attempt to address this problem by conducting cardiac screenings on elementary school children in Texas. If this statewide program is successful, similar cardiac screenings could be mandated in more states to identify children at risk for SCA.

The Championship Hearts Foundation (CHF) was awarded a 2-year, $1-million contract by the Senate to carry out the pilot program. The CHF is an Austin based nonprofit that has organized more than 6,000 heart screenings for student athletes. The contract brings together the collective expertise of the CHF, Children’s Medical Center of Dallas, Texas Children’s Hospital of Houston and Children’s Cardiology of Austin.

Under the proposed pilot program, licensed technicians will provide free screening to 12,750 sixth grade students in an attempt to identify any conditions that could lead to heart failure. The students will fill out a pre-screening questionnaire and receive an electrocardiogram and echocardiogram. Board-certified/approved cardiologists will read and interpret the tests and then send the results to the parents. Once all the screenings are conducted, the results will be forwarded to the Texas Education Agency by June 2009 to decide if a larger, statewide screening program should be implemented.

Software Update from Medtronic Identifies Potential Defibrillator Lead Fractures

In September, the U.S. Food and Drug Administration (FDA) approved a software update from Medtronic that helps detect fractures of the company’s Sprint Fidelis implantable defibrillator lead. Called the Lead Integrity Alert, the new software modifies the device settings so the defibrillator can better detect whether a lead has fractured or an abnormal heart rhythm has occurred. Once a lead fracture has been determined, it issues an audible alert which repeats every four hours until a physician can reset the defibrillator. This software update will reduce the occurrence of inappropriate defibrillator shocks and ensure that the device is working properly.

The Sprint Fidelis lead, the subject of an October 2007 Medtronic recall, was prone to fracture in a small number of patients. Most patients with the Sprint Fidelis lead still have the device implanted because of the surgical risk associated with removal, and are closely monitored by their physicians for potential fracture.

Genetic Information Nondiscrimination Act Passed

In May 2008, President Bush signed the Genetic Information Nondiscrimination Act (GINA) into law. It is the first major civil rights bill of this century. GINA provides individuals with strong, enforceable federal protections against the misuse of genetic information in health insurance coverage and employment decisions. As a result, some of the major concerns of families considering genetic testing are removed.

The health insurance provisions of the bill, Title I, will take effect in May 2009. It guarantees that insurers cannot request, use or obtain genetic information or genetic tests from an individual or a family member to determine eligibility, coverage or premium-settings. The protections in employment, Title II, will take effect in November 2009. It guarantees that, except in rare cases, employers, employment agencies, labor organizations and training programs may not request, use or obtain genetic information from an individual or family member to make decisions about hiring, firing, training, compensation, promotion or membership.

We Want to Hear from You!

Let us know what you would like covered in future newsletters. We encourage parents, relatives, physicians and nurses to submit items for consideration. Articles can be a maximum of two pages and sent as an attached MS Word document to newsletter@childrenscardiomyopathy.org. Photographs or artwork should be submitted as image files (.tif, .gif, .jpeg). The deadline for our Spring/Summer 2009 issue is March 16th.
**For Families**

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**Q&A with David R. DeMaso, M.D.**

Helping Children Adjust to their Diagnosis

Dr. David DeMaso is a Professor of Psychiatry and Pediatrics at Harvard Medical School and Psychiatrist-in-Chief and Chairman of Psychiatry at Children's Hospital Boston. As a child and adolescent psychiatrist, the underlying essence of his work is to understand what facilitates or hinders an individual’s ability to cope with adversity. For the past 30 years, he has worked in the Department of Cardiology with the pediatric heart transplantation team in particular.

Adjusting to a life without competitive sports, especially if they excelled in sports or come from a sports-oriented environment can be quite difficult for a pre-teen or teen. How can a parent help them deal with the shift in identify from “athlete” to “non-athlete” in a positive way?

With any loss comes grief. Grief is accompanied by sadness and anger. It begins with a period of shock and even numbness, but within days to a couple of months feelings of protest and anguish come to the surface. The diagnosis of cardiomyopathy and accompanying limitations represent a significant loss for any child. Parents can help by acknowledging, understanding and accepting these normal emotional reactions that will undoubtedly arise within their child following diagnosis.

While difficult, it is important to listen to these feelings. Naturally no one wants his or her child to be in pain. Parents want to come up with solutions that will take the hurt away. However, for a child to move forward, it is important and helpful for him or her to feel heard and to have their disappointment accepted. Listening allows the expression of loss, minimizes feelings ofaloneness and allows a child to mourn their loss. While painful, mourning allows a child to move to a new view of him or herself. It can allow the child to accept “what they can do” and “what they cannot do.” Listening allows a child to hear about the “other things that they are good at” and the “other things in life that are still available.” Listening is doing something.

When heard, a child is more receptive to hearing parental advice and encouragement to pursue new activities or interests. These suggestions can range from photography to art to music interests. One can still be a sports fan or follow sports as a manager or scorekeeper. The point is that by accepting and understanding a child’s frustration and disappointment, a parent can help a child “let go of the past” and develop a new sense of self.

Many children with cardiomyopathy once they hit the pre-teen or teen years are angry or resentful of their diagnosis. Some may outright blame their parents for passing the disease to them. How can parents help their child view things more positively and find an outlet for their anger and frustrations?

This is part of grieving. Anger at one’s parents is a natural reaction for a child who ideally would like to think that his or her parents can and should protect them from any hurt or disease. The best response to this anger is for a parent not to be defensive. Being open to a child’s anger and resentfulness will, over time, prove helpful in resolving their upset and arriving at a new view of themselves in relation to their illness. Remember; a child is actually upset at their illness and limitations and less so at the parent per se. Becoming defensive can only add something else for the child to be upset about.

Some teens may refuse to accept their condition and decide not to take their medication or do the medical tests recommended by their cardiologist. What can parents say or do to make them realize the seriousness of their diagnosis and be proactive and responsible for their heart health?

This is a very difficult problem for some youngsters. A parent can begin to approach this problem by making sure that the teen has a full understanding of their illness. In the midst of so many feelings generated by the illness, one might actually find that the teen does not really understand their illness and its treatment. The teen may have significant misinterpretations that interfere with his or her care plan. Often together with the cardiology team, a teen’s understanding of his or her illness can be reviewed and lead to helpful re-education of the teen with an increase in adherence.

From the beginning, it is helpful to simplify the treatment regimen whenever possible. Helpful strategies to remind patients about their treatment include the use of pagers, alarm clocks and telephone calls from parents or medical clinics. Other strategies include using pillboxes, storing medications in highly visible places and posting reminders around the house. The first step in treating nonadherence when it is apparent is to increase the level of supervision. This increase may involve parental observation or administration of treatments, more frequent clinic visits or laboratory monitoring. The use of incentives tends to be more effective than educational or organizational strategies alone. For younger children, a program may involve a sticker chart tied to age appropriate incentives. For
adolescences, adherence may be tracked using signatures on a chart with a similar system of short and long-term incentives. If the above steps do not resolve the problem, it becomes important to make sure that the grief process described above has not evolved into disabling depression and/or anxiety. The teen that becomes clinically depressed or anxious generally also struggles with other areas of his or her life such as academic functioning or school attendance. Teens that have had pre-existing difficulties with depression, anxiety or oppositional behavior may be at particular risk for troubles adhering to their medical regimen. The teen struggling with these kinds of problems can benefit from a psychiatric evaluation focused upon assessing their strengths and vulnerabilities as well as developing new coping strategies.

Some teens have mentioned that certain classmates no longer felt comfortable spending time with them because of the possibility of a cardiac emergency or because they no longer have a common interest (e.g. sports). How can a parent help their teen feel less isolated in this difficult situation?

Teens indeed may report that friends and even the school are afraid. The cardiology team can work with the teen and parents to provide the necessary education for the friends and schoolmates regarding the illness. Information can be critical in reducing these fears.

In some situations, a child’s classroom can benefit from information regarding the illness, particularly if the child needs to be hospitalized during the school year: Providing information to the classroom should be done only if the child wants this to occur. Parental permission and review of “what will be said” is critical. This recommendation is generally most useful for younger children. Older children tend not to want to highlight differences from their peers, particularly in the classroom.

What should a parent do if they notice their teen becoming withdrawn and socializing less with friends because of their heart condition?

Parents should not hesitate to bring up their observation to their teen. Based on a teen’s response, parents might support and even encourage their teen to make contact with their friends. If more concern seems to arise the parents might consider obtaining a psychiatric assessment, which can be phrased as getting an assessment for the “whole family…You have an illness that affects all of us…We plan to meet with someone to help us cope with this new event.” This takes the focus off the youngster as the “cause of the problem.”

What steps should a parent take if they observe signs of depression in their child?

The classic warning signs of a clinical depression are the presence of persistent depressed mood or markedly diminished interest or ability to have pleasure. There

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A Different Drummer: A Mother’s View on HCM and Depression

By Mary W.

Two years ago, after my son Jake was diagnosed with hypertrophic cardiomyopathy, he fell into a deep depression. He was ten years old at the time.

Jake has always been a physical kid. Before his diagnosis, he loved to run and especially to sprint. He excelled in gymnastics, and his life’s dream was to become a stuntman in Hollywood. Suddenly, Jake needed to limit the risky acrobatic stunts that he loved, to cut back on his rock-climbing, and to stop sprinting altogether. How do you tell a kid who loves to run like the wind that his new form of exercise will be riding his bike slowly and steadily on a flat surface?

Although Jake initially insisted that he was fine, gradually he began to change. He continuously wore the hood of his sweatshirt up over his head regardless of the weather. He even slept with that hood on, crawling into bed in the clothes he had worn during the day. His grades plummeted, and we started getting phone calls and emails from his middle school. “Jake has his head down on the desk all through class,” his math teacher wrote. “Jake isn’t participating at all,” wrote his social studies teacher. “He sits in the back of the room and stares at the wall.”

Jake began to talk about wanting to die. I put up a white board in his room and every night he drew a version of the same scenario in which a little angel was destroyed in some violent way by a huge evil monster. When we asked “Why does the angel always have to die?” Jake responded “Because there’s no other way out.”

We were at a loss as parents. Upon the recommendation of the school psychologist, we met with an expert on adolescent depression and poured out our hearts. He said, “Of course Jake can’t concentrate on school right now. He’s coming to terms with his own mortality. What adult in his position would be able to go to school, do homework, sit in a desk listening to lectures?” I wanted to throw my arms around him; I was so grateful for his words. He suggested that we take some of the school pressures off of Jake. He also recommended that we draw up a 504 plan with the school, outlining the restrictions and psychological factors that are part of HCM. This way the school would be required by law to make some accommodations for his depression.

We all began to relax about school and let Jake deal with his diagnosis the way he needed to, whether that meant withdrawing for a while, getting angry, or creating art that was a little scary to the rest of us. His depression began to lift. Now he plays the drums. Through music, he expresses himself, pounds out some energy and gets the rest of us dancing.
Relevance of STATE WAIVER PROGRAMS for Children with Cardiomyopathy

When a child is diagnosed with cardiomyopathy, there may be certain out-of-pocket expenses for treatments and medications that are not covered by health insurance. For children with more severe cases of cardiomyopathy, waiver programs could assist in covering costs such as medicines, therapies, nutritional supplements or care in the home from a nurse or home health aide. Parents of an affected child are encouraged to investigate the waiver program, as well as other state programs, to see if their child qualifies for financial assistance.

Waiver programs are devised to help children with disabilities remain in their own homes instead of living in institutions. Funding for these state-run waiver programs comes from both state and federal sources. When a child is on a waiver program, the state “waives”, or does not count, the parents’ income when assessing if the child meets the program’s financial criteria. When a child is accepted to a waiver program he or she receives Medicaid benefits that can help cover some of the costs associated with care at home.

While not every state has a waiver program, specific benefits and eligibility criteria vary among those that do. Many states require a child to be considered disabled in order to receive waiver services. A diagnosis of cardiomyopathy alone is not enough to determine if a child meets the criteria for being disabled. Factors considered include the severity of the cardiomyopathy and the child’s ability to perform normal daily tasks. Waiting lists can be long so it is advisable for families to find out what is available in their state as soon as possible.

To find the correct state office to contact, parents can do an internet search for “Medicaid”, “waiver”, with the name of the state to determine the program name. Waiver programs for children have many names - the Katie Beckett Waiver, Deeming Waiver, Medicaid waiver, 2176 Model Waiver, and more. No matter what the waiver programs are called, they can be an important resource for parents who care for a chronically ill child at home.

Q&A: Helping Children Adjust

continued from page 9

Can be significant appetite, weight, and sleep changes that go beyond those directly attributable to the teen’s heart condition. There may be troubling feelings of worthlessness or guilt as well as diminished ability to concentrate. Recurrent thoughts of death or even suicidal ideas may be present. These symptoms are present most of the day or nearly every day for longer than a 2-week period. Parents should begin by letting their child know about their concerns as well as letting the child’s primary care clinician and/or cardiology team know about their observations. Psychiatry assessment should be sought when these symptoms are disabling, troubling or puzzling. The primary care clinician and/or cardiology team can be helpful in determining where to find this assessment. The child can be told that they are getting a “mental health check-up” just as they get a “physical check-up.”

If a parent suspects that their child is suicidal, Parents should seek immediate psychiatric evaluation. The child’s primary care clinician and/or cardiology team can be contacted to arrange for this assessment. A child can also be taken to the nearest emergency room for an acute psychiatry assessment.

Is there a specific type of psychologist, therapist, or counselor that a family or child with cardiomyopathy should see?

Ideally it is helpful for a child and family to see a mental health clinician familiar with cardiomyopathy. Needless to say, this is usually not available. However, parents can ask for a mental health clinician’s familiarity in working with acute and chronic physical illnesses. Many of the issues involved with cardiac illness have parallels that cut across different pediatric physical illnesses.

Are there any resources that you can recommend for teens with chronic illness?

With colleagues and families, we have developed the Experience Journals at Children’s Hospital Boston (www.experiencejournal.com). These journals are designed to promote the healthy coping of children and their families who must contend with significant physical and emotional illnesses. The Experience Journals are collections of stories, pictures and personal experiences from families about what it has been like to live with their children’s illnesses. They represent the “collective wisdom” of these children and their parents as well as their health care providers. We welcome family members to enter and explore any or all of the Experience Journals.

For those children who have been transplanted, they face different challenges; their medications may cause excessive weight gain, severe acne and/or more facial hair. Then there is visible scarring from the heart surgery itself. Given all this, what kind of support can a parent give to a more image-conscious teen after their transplant?

My research in adapting to cardiac illnesses has repeatedly pointed out that “how a child does emotionally” correlates more with “family functioning” than with the severity of a child’s heart condition. I have never been able to reliably predict “how a child is emotionally” based upon the medical status of his or her physical condition. I have seen acutely ill children who are emotionally quite well. And conversely, I have seen physically well children, who are emotionally quite troubled. The difference often lies in a supportive and attuned family. Thus, my recommendations for listening to children and understanding the disease are grounded upon increasing family support. A child’s resiliency and adaptation to adversity are greatly enhanced by a family that listens and understands.
The Effect of Passing on a Gene: Grief, Guilt and Dedication

By Heather Skirton
PhD, Registered Nurse and Genetic Counsellor (UK)

Health outcomes for affected children, but there seems to be little research on how a family can actually live with a chronic heart condition. While the search for cures or better treatment goes on, families still have to live and survive the day-to-day physical and psychological pressures of having a child with a serious condition. The impact is enormous whatever the situation, but when the condition is inherited, there may be additional feelings of grief and guilt on the part of the parent(s).

Even though passing on our genes is something we have absolutely no control over, many parents still feel guilty. When told her two year old son’s condition could have been the result of a gene mutation passed on by her, one mother responded, “It made me feel as though I was to blame, that my husband shouldn’t even be with me, because I was the one that carried the child for nine months, gave birth to it and brought out one that wasn’t right”. Yet another parent said, “I do blame myself, I blame myself the way he is, that it’s something I’ve done wrong”. However, when asked, most teenage or adult children say that they realize it is “no-one’s fault” and they do not wish their parents to feel guilty.

This guilt is not confined to mothers; fathers also feel this way but may find it more difficult to express and have less access to social support from friends and colleagues than women do. Despite changes in the structure of our society with respect to roles, men still feel pressure to “stay strong” for their partners and children, often at considerable emotional cost to themselves.

While there is little evidence of one parent actually blaming the other, parents who have passed on the mutation often say they are fearful that the other parent blames them for what has happened. Feelings of guilt and despair can create discord between partners, if not discussed. Talking about your feelings with experienced professionals can be very helpful. A couple who had lost two children several years ago talked about the guilt they felt with a counselor. Both felt it would upset the other parent if they raised the subject, but being with a third ‘neutral’ person enabled them to talk to each other. They had been overwhelmed by grief and guilt that they had caused their children to suffer, but found after several sessions with the counselor that they were able to carry on speaking about their children to each other in a more natural and comforting way.

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Many parents also find that talking to parents who are in similar situations (e.g. through a support group) benefits them enormously, as they feel that their experiences and feelings can be truly understood. Finally, it sometimes helps to put things into perspective by remembering that none of us can promise our children perfect lives. We can only strive to support our children and fulfill their needs to the best of our abilities.

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