From the PRESIDENT

It’s hard to believe that another year is ending. With a blink of the eye, 2006 will soon be over! I find that often it gets so busy during the year, I forget to sit back and enjoy all that has been accomplished. In working on this issue, it was therefore rewarding to read about the many new projects in development and how CCF supporters and families continue to be involved.

Recently, Pauline and I attended a Medtronic sponsored sudden cardiac arrest meeting, which encouraged us to reflect upon CCF’s purpose and achievements. We had the opportunity to connect with other patient heart organizations and discuss commonalities from a broader perspective. Taking an aerial view of issues from “50,000 feet” made us realize that many of the other groups are trying to achieve the same thing as us – prevent premature loss of life. The meeting was a reminder to come together to achieve more as a cohesive group. This concept also applies to CCF’s situation – how can we as donors, families, and medical professionals work together to help more children affected by cardiomyopathy? It is something worth thinking about, especially at this charitable time of the year.

Even in the midst of the holiday rush, I hope that you will find time for positive reflection, which includes acknowledging yourself for a job well done. This means, for medical professionals, your commitment to caring for children with cardiomyopathy; for families, your perseverance in searching for answers to your child’s condition; and for donors, your compassion for wanting to make a difference.

May the holidays be especially meaningful and rewarding for you and your family.

Lisa Yue  
Founder & President

CCF to Fund New Genetic Research on DCM & HCM

A critical issue with pediatric cardiomyopathy is that the majority of diagnosed children do not have a known cause for the disease. According to the Pediatric Cardiomyopathy Registry, less than 25% of all patients have a defined etiology despite rigorous, standardized evaluation. There is less of an understanding of the causes of the disease in children because there are fewer affected patients and therefore less clinical data and patient samples available for research. CCF is trying to address this problem by supporting investigators who have an interest in researching pediatric cardiomyopathy and have access to a larger cross section of patient data and samples. Earlier this year, CCF approached Jeff Towbin, MD of Baylor College of Medicine about conducting additional molecular research on pediatric cardiomyopathy. Dr. Towbin is the Chief of Pediatric Cardiology at Texas Children’s Hospital and a Professor of Pediatrics at the Baylor College of Medicine. A well-known leader in the field of pediatric cardiomyopathy, Dr. Towbin manages the largest cardiomyopathy clinic and service for children in North America, seeing close to 1,000 patients annually. Over the course of a year, CCF will award $92,250 towards Dr. Towbin’s study, Identification of Mutations.

SCIENTIFIC PAPERS PUBLISHED with CCF Support

Three important scientific papers are being published that will provide better understanding of pediatric cardiomyopathy and hopefully will lead to more effective treatments for children with the disease. Partnering with the National Institutes of Health funded Pediatric Cardiomyopathy Registry (PCMR), CCF sponsored think tank sessions that enabled the analysis of the registry’s patient data from 252 medical centers. Findings from the working group were initially presented at the American Heart Association 2004 Scientific Sessions. Now, final manuscripts will be made available to medical professionals world-wide in three respected scientific journals. Publications in journals like the Journal of American Medical Association undergo a rigorous peer review process, and they generally accept less than 1% of submitted articles.

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A Cause for Today...A Cure for Tomorrow
The three articles acknowledging CCF’s support are:

**Factors Associated with Establishing a Causal Diagnosis for Children With Cardiomyopathy** (Gerald Cox, Lynn Sleeper; April Lowe et al.) was recently published in the October 2006 issue of the journal *Pediatrics*, the official journal of the American Academy of Pediatrics. The goal of the study was to identify the clinical factors associated with determining a cause of cardiomyopathy in children. In studying the PCMR findings, researchers discovered that only one third of the 916 patients with pediatric cardiomyopathy had a known cause at diagnosis. “We believe one of the main reasons for the poor outcomes is that in the majority of affected children we do not know the cause, so everyone is being treated the same way, instead of tailoring a treatment to fit the cause,” commented Steve Lipshultz, MD, principal investigator of the PCMR and the head of CCF’s medical advisory board.

In an effort to standardize diagnostic approach, the study recommends that the evaluation of cardiomyopathy in children include a thorough review of family history, echocardiograms for first-degree relatives, and routine blood and urine testing to pick up such things as inborn errors of metabolism, neuromuscular disorders and viral myocarditis. If a cause is still not found, endomyocardial biopsy for children with dilated cardiomyopathy should be considered, and for children with hypertrophic cardiomyopathy where a mitochondrial disorder is suspected, a skeletal muscle biopsy should be performed.

**Incidence, Causes and Outcomes of Dilated Cardiomyopathy In Children: Findings from the Pediatric Cardiomyopathy Registry** (Jeffrey Towbin, April Lowe, Steve Colan et al.) was also published in the October 2006 edition of the Journal of American Medical Association. This paper sets out to provide a detailed description of the incidences, causes, outcomes, and related risk factors for dilated cardiomyopathy (DCM) in children. Several key factors emerged from the 1,426 DCM patients that they studied: more boys are affected than girls, blacks more than whites, and infants more often than children. The most common known causes were myocarditis (heart inflammation) and neuromuscular disease, but the majority (66%) of cases had no known cause. The study shows that knowing the cause of cardiomyopathy can predict how a child will subsequently do. It also reveals that children with certain types of DCM may not be transplanted as often as they should. With the study findings, pediatric cardiologists will now have better risk stratification information to make more accurate decisions about when to list a child for cardiac transplantation.

The third manuscript, *Epidemiology and Etiology-Specific Outcome of Hypertrophic Cardiomyopathy in Children: Findings from the Pediatric Cardiomyopathy Registry* (Steve Colan, Steve Lipshultz, April Lowe et al.) has also been accepted for publication in a leading peer review journal. The study identifies the various causes for HCM in children and determines the relationship between outcomes, etiology, and age at presentation. Key findings from this study will be revealed after publication in the next few months.

These publications are important in guiding other physicians caring for children with cardiomyopathy. “New methods for diagnosis, new therapies, and a better understanding of the characteristics of all patients will hopefully lead us to better outcomes for these children in the future,” adds Dr. Lipshultz.

Original publications can be accessed through www.pubmed.gov.
NEW Patient Booklet  Available FOR CHILDREN

CCF and the National Society of Genetic Counselors (NSGC) have partnered to develop a new patient publication entitled Cardio What? - A Kid’s Guide to Cardiomyopathy.

Written by genetic counselors from Johns Hopkins, University of Colorado at Denver, and the University of Chicago, the booklet teaches at risk and diagnosed children with cardiomyopathy the basics about the disease in an interesting and informative manner. The publication is meant to be complementary to CCF’s Understanding Pediatric Cardiomyopathy booklet developed for parents and caregivers.

Targeted to children 8-13 years old, the 26-page booklet uses simple child-friendly language, colorful illustrations, and fun word games to help children better understand their own and/or their family member’s diagnosis of cardiomyopathy. Although intended for children affected by cardiomyopathy, it can also be shared with siblings, relatives, and friends to explain their inherited condition.

Topics covered include: how the heart works, an explanation of cardiomyopathy, common symptoms, causes of the disease, overview of genetic inheritance, associated tests/procedures, taking care of oneself, and an explanation of how cardiomyopathy can affect a child’s life and emotions. Puzzles, word searches, fill-in-the blanks, coloring, and animated pictures help to make this complex disease more easily understood and less frightening.

The idea for the booklet came from the realization that most children affected by cardiomyopathy do not fully understand the disease, the purpose of screening examinations and genetic testing, or what it means to live with the disease. In trying to explain the disease, NSGC members found that there was no age-appropriate literature developed for children with familial forms of cardiomyopathy. At the same time, CCF was interested in developing a booklet that could empower young children with cardiomyopathy to educate themselves and proactively manage their disease. CCF also wanted to stress to physicians and family members the importance of working with a geneticist and/or genetic counselor. From this mutual interest, both parties decided to collaborate in research, development, and funding of the booklet.

The booklet will be distributed to pediatric cardiologists, nurses, social workers, geneticists, and genetic counselors as part of a series of patient education materials on pediatric cardiomyopathy. CCF and NSGC will be notifying hospitals of this new resource through direct mail and medical meetings in the coming months. For a limited time period, CCF will provide booklets free of charge to interested physicians and families. To place an order, please contact Stormy Hill at thill@childrenscardiomyopathy.org with your order quantity and mailing address.

NEW PATIENT EDUCATION DVD IN DEVELOPMENT

A key finding that emerged from past research was that in terms of patient education materials, parents preferred technical information about the disease such as terminology, symptoms, testing, and treatment options to be balanced with practical information on quality of life and coping issues. Several doctors suggested that a patient DVD might be a good way for newly diagnosed families to connect emotionally with others in a similar situation and to provide the much needed hope and encouragement to deal with the daily challenges and uncertainties of living with a chronic disease.

With grants from the American Legion Child Welfare Foundation and eBay Foundation, CCF has started working on a DVD designed to provide the kind of information that parents and physicians are seeking. The 20-minute DVD entitled Secrets of the Heart - Living with Pediatric Cardiomyopathy will cover topics such as: what is cardiomyopathy and how did my child get it; what is the typical treatment; how does this affect our daily life (physical/psychological issues, school, friends, etc); and what is the best way to cope with the disease?

Filming recently took place in New York City and Houston, Texas. The DVD will feature interviews from four physicians with expertise in pediatric cardiomyopathy and four families whose children were diagnosed with either hypertrophic, dilated, or restrictive cardiomyopathy. The DVD will highlight their experience of living with cardiomyopathy from both the parent and child’s perspective and cover the common concerns about the disease. Currently the DVD is in the post-production, and plans are in place to have the DVD ready for distribution by January 2007.

The DVD is being developed under the creative direction of Lindsay Anderson of 44Percent Design in Chicago. Lindsay will oversee production, working alongside Ruth Krueger, Copywriter/Creative Strategist and Andreas Larsson, Director of Photography. Members of the creative team have collectively worked for many well recognized clients such as Caterpillar, McDonald’s, Philips, Nestle, Abbott, Eli Lilly, and CBS to name a few.
In 2006, CCF embarked on a year long Patient Outreach Initiative to more effectively reach and support families of children with cardiomyopathy.

Funded in part by a Medtronic Patient Link grant, the goals of the initiative are to service more cardiomyopathy families with appropriate information and resources, offer more local support services, and enhance CCF’s relationship with the top cardiomyopathy physicians and medical centers.

To lead this Patient Outreach Initiative, CCF created a new part time patient outreach and support position. Filling this new coordinator role is Dr. Talitha “Stormy” Hill who joined CCF in March 2006. Stormy is no stranger to CCF. She has been part of the CCF family network since 2004. Stormy’s son has dilated cardiomyopathy and when he was diagnosed, a social worker informed Stormy about CCF.

As the new CCF Patient Outreach and Support Coordinator, Stormy will be responsible for creating, overseeing, and improving CCF’s network of hospitals and families. Her position will focus on four main areas: 1) improving patient support including the family registration process, 2) strengthening and expanding CCF’s hospital relationships, 3) increasing family registrations through outreach initiatives, and 4) establishing and managing local support groups.

Stormy graduated from the College of Human Medicine at Michigan State University this Spring and has a special interest in child adolescent psychiatry with emphasis on the psychosocial issues that arise in children with chronic illnesses. In the past, Stormy conducted research on the psychosocial stressors related to raising children with heart disease and created a parent resource guide.

Stormy always had an interest in working in the non-profit field. “With my background, it is exciting to be able to work at a non-profit and help a foundation that deals with cardiomyopathy,” Stormy says. Stormy’s medical background allows her to understand the medical jargon involved and her experience as a mother of a child with cardiomyopathy allows her to put herself in the shoes of other parents in a similar situation. Lisa Yue agrees that CCF’s position is a natural fit for Stormy. “We are thrilled to have Stormy join us. Her background is perfect for what we are trying to accomplish, and we feel fortunate to have found such an enthusiastic and motivated person for the job,” she says.

A Children’s Booklet about Special Hearts

Daren and Stormy Hill are the proud parents of Mason. Mason is 2 1/2 years old and is diagnosed with dilated cardiomyopathy/left ventricular non-compaction related to a mitochondrial disorder. In honor of their son, they produced a children’s booklet, Mighty Mason’s Special Heart. Written by Mason’s dad and illustrated by Mason’s grandmother, the 16-page fun and easy-to-read booklet is ideal for children ages 2 to 6.

The inspiration for the booklet came from the Hill’s own experience of dealing with Mason’s diagnosis. Due to his mitochondrial condition and heart disease, he struggles with many health issues but he continues to beat the odds. “To look at him you would never imagine that his heart is diseased as he is so energetic and full of life,” says Stormy. When he was hospitalized for heart failure his dad nick-named him “Mighty Mason” for his fighting spirit and the name has stuck with him ever since. Initially Daren and Stormy created the booklet so that Mason would have something new to read but they soon realized that Mason’s story could be an inspiration to other “heart kids” and their families.

The colorful booklets are now being sold for $4 each (includes shipping) and 50% of the proceeds will be donated to CCF. To read Mason’s story and to order the booklet, please visit www.specialhearts.net for more information.
One of CCF’s key initiatives for 2006 has been to provide more local support services to better meet the informational and emotional needs of diagnosed families. Establishing local support groups has achieved even greater importance, since CCF’s network of families has grown from 10 to 450 families in the past four years. CCF realizes that face-to-face interaction is important for members to share experiences, learn from one another, and provide hope and support to each other.

Currently, there are two support groups established (Georgia and Michigan) and four more are in the formation stages (Tennessee, Massachusetts, North Carolina, Idaho). All six are volunteer-led by CCF family members. CCF works closely with each local support group leader to involve hospital support staff to promote and facilitate the meetings. Partnering centers help to mail out introductory and/or meeting letters to their database of cardiomyopathy patients. This has been key to reaching new families and getting existing families more involved.

The Georgia group was the first local support group to be established in early 2005. Led by Audrey Callahan, the group works with the Sibley Heart Center to do patient mailings and organize meetings. Shannon Chapman, Program Specialist with Kids at Heart, and Alex Berg, RN and Transplant Coordinator; help facilitate the meetings. Dr. William Mahle serves as the group’s medical consultant. The Georgia group has had three meetings and three informal get-togethers thus far. Occasionally, Audrey is also called in to speak to cardiomyopathy families admitted to the hospital. The group’s activities include participating in the American Heart Association Heart Walk in November, organizing a Q&A with a pediatric cardiologist in January, and planning a fundraiser in June 2007. This past August, Audrey spoke about CCF and her experience with pediatric cardiomyopathy at the Fourth Annual Centers for Disease Control and Prevention (CDC) conference. She was part of a family panel discussion on “Faces of Congenital Heart Disease” and presented to 80 doctors, nurses, CDC representatives, and researchers.

The Michigan group is led by Brian Hill. Since there are two primary medical centers that service southeast Michigan, this group is partnering with both C.S. Mott Children’s Hospital and Detroit Children’s Hospital. Dr. Robert Ross and Dr. Robert Gajarski are the medical advisors and Jill Matson, RN, and Meg Zamberlan, RN and Transplant Coordinator; serve as the medical support staff. Introductory letters have been sent to 150 cardiomyopathy families, which have tripled annual family registrations for this area in a 3 month time period. The first meeting was held November 19th, and plans are to have one meeting per quarter with two held in Ann Arbor and two held in Detroit.

The North Carolina group led by Angela Henderson is hoping to partner with Duke University Medical Center. A social worker and two nurses have volunteered to assist the group. Jinjue Serre leads the Idaho group and is looking to partner with St. Luke’s Medical Center. Massachusetts leader, Stephanie Thorsen, is in the process of contacting Children’s Hospital of Boston and reaching out to registered families in Massachusetts and the surrounding New England states. Tennessee leader, Patricia Simpson, is also in the process of contacting Vanderbilt University to host group meetings.

A new support group manual has been developed to assist other CCF members interested in starting a local support group. This manual will take leaders through the steps to get a local group established in their state. The manual includes sample letters to hospitals and families, worksheets, planning guidelines, and a sample meeting agenda.

For additional information, please contact Stormy Hill, Patient Outreach & Support Coordinator at thill@childrenscardiomyopathy.org or 886-808-CURE, ext 905.
CCF to Fund New Genetic Research on DCM & HCM

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in Genes Associated with Hypertrophic Cardiomyopathy and Dilated Cardiomyopathy. This study will be complementary to Dr. Gerald Cox’s study, Analysis of Sarcomere Gene Mutations in Pediatric Hypertrophic Cardiomyopathy, that CCF funded in 2005. As part of CCF’s funding requirement, both investigators have been asked to share research data in a collaborative manner for their publications.

The two key objectives of this research is to 1) identify the genes responsible for the clinical phenotypes in children with cardiomyopathies and 2) define the causative mechanisms involved in the development and maintenance of childhood cardiomyopathy. In this new study, Dr. Towbin will perform a comprehensive genetic screening of the major genes associated with dilated cardiomyopathy (DCM) and hypertrophic cardiomyopathy (HCM). DCM and HCM are the two most common forms of cardiomyopathy, accounting for 85% of the cases in diagnosed children. Using DNA sequencing, two hundred children will be screened for the 11 most common HCM genes and the 9 most common DCM genes.

While the premise for this study is simple, it has not been done in previous years due to the lack of quality samples and funding. These obstacles will not, however, be an issue with Dr. Towbin’s study. Dr. Towbin directs one of the top cardiac molecular genetics laboratories in the country. His lab has one of the largest reserves of patient data on the disease with over 800 DNA samples from patients with cardiomyopathies, including familial and sporadic cases. Now that the right resources are in place, CCF is eager to get this important study started. “Because Dr. Towbin already has a large number of samples, we knew that with the appropriate funding, he would be in the best position to find answers to the causes of the disease,” says Lisa Yue, CCF’s Executive Director.

Dr. Towbin has devoted most of his medical career to understanding cardiomyopathy in children. He began studying the molecular basis of X-linked DCM in 1989 and his research led to the discovery of dystrophin, a gene responsible for DCM. Since 1989, four genes causing DCM have been identified in his lab. At the time, he also noticed that no one was taking an interest in caring for pediatric cardiomyopathy patients. “I was interested in the genetics of cardiomyopathy, and I thought that, while I am doing research on these patients, I can take care of them as well,” says Dr. Towbin.

The new proposed study is significant because it will provide critical data on the genetic makeup of children with cardiomyopathy. Most work to date has focused on identifying genes in adults and then seeing if children carry the same gene. The novelty of this study is that it will explore it from the children’s standpoint first. By obtaining cross-sectional data that identifies the type and location of mutations associated with pediatric cardiomyopathy, the study will determine the applicability of adult data to pediatric patients, identify appropriate direction for new molecular work in children, and clarify the genotype-phenotype relationship. The study will also help physicians understand the role of genetic testing in patients with cardiomyopathy and provide guidelines for testing based on patient-specific characteristics. For Dr. Towbin, the great expectation for this study is that “we will find some new genes in this study and identify a number of causes of pediatric cardiomyopathy, specifically for dilated cardiomyopathy and hypertrophic cardiomyopathy.”

A Note to Medical Professionals

If you or your center would like additional copies of the Heart to Heart Newsletter for patient or physician distribution, please email your request to CCF at newsletter@childrenscardiomyopathy.org and indicate your mailing address and number of copies desired.

CCF Attends SUDDEN CARDIAC ARREST Meeting

Sudden cardiac arrest (SCA) is a major health problem. According to the American Heart Association, SCA kills more than 45,000 people each year in the United States. Many of the victims are children with inherited heart rhythm disorders such as arrhythmogenic right ventricular cardiomyopathy or hypertrophic cardiomyopathy.

CCF was recently invited to participate in a Medtronic sponsored meeting, Sudden Cardiac Arrest Connect. Held October 24th at Medtronic’s World Headquarters in Minneapolis, Minnesota, seven patient organizations came together to discuss ways to prevent and treat sudden cardiac arrest. Other participating organizations included the Cardiac Arrhythmias Research and Education Foundation, Hypertrophic Cardiomyopathy Association, Mended Hearts, Parent Heart Watch, Sudden Arrhythmia Death Syndrome, and Sudden Cardiac Arrest Association.

A professional strategic planner facilitated the brainstorming session and guided the discussions on possible collaborations in the area of national awareness and primary prevention. Members of the group identified a subcommittee who will draft a concept outline and funding proposal for a collaborative SCA public awareness campaign.
Pediatric cardiomyopathy is a complex and extremely variable disease, which requires a thorough evaluation and careful monitoring. When a child is at risk or diagnosed with cardiomyopathy, a full investigation should take place at a center with the specialized resources to perform a comprehensive medical, genetic, and surgical evaluation. Many families contact CCF to locate a physician with expertise in pediatric cardiomyopathy at a specialty center or clinic.

A pediatric cardiomyopathy specialist is defined as a physician who focuses on patient care or research in the fields of cardiomyopathy, congestive heart failure, electrophysiology, heart transplantation, and/or the genetics of cardiomyopathy. A specialist may be affiliated with a pediatric cardiomyopathy clinic or comprehensive care program. There are only a handful of centers in the U.S. and Canada that have a specialized clinic or program for cardiomyopathy patients. Usually they take a multi-disciplinary approach to evaluation, treatment, and long-term care, and there is coordination among pediatric cardiologists, geneticists, nurses, electrophysiologists, pediatric heart surgeons, nutritionists, pediatric neurologists, and social workers.

To ensure that affected families receive the most advanced care, CCF is updating its physician and hospital list of the top cardiomyopathy and heart failure centers in the U.S. and Canada. In December, CCF will mail hospital surveys to 280 centers across the country to collect information on various pediatric heart programs. The survey will verify the resources available at each center, specify the physicians with a clinical interest in cardiomyopathy or in related fields, and provide clinic dates and times specific to their cardiomyopathy practice. Centers will also be asked to give input into how CCF can better serve the needs of cardiomyopathy patients and their families.

From the completed center surveys, a specialist list will be developed which will include specialty centers and physicians searchable by city and state. Registered families can then access this information from CCF’s website starting February 2007. This will be an important resource for helping families find physicians and centers with the most experience in treating children with cardiomyopathy.

If your center or clinic received a survey, we encourage you to fill it out as soon as possible and return it via fax or the supplied postage paid envelope. If your center or clinic did not receive a survey and would like to be included on the specialist list, please contact Stormy Hill, Patient Outreach & Support Coordinator at 866-808-CURE, ext. 905 or thill@childrenscardiomyopathy.org.
Children and Implantable Cardiac Defibrillators: What Every Parent Should Know

Most everyone has seen someone on television holding paddles and heard the actor call out “Clear” before they place the paddles on the patient to try to shock their patient’s heart back into a normal rhythm.

While this may appear overly dramatic, the same kind of technology applies to implantable cardiac defibrillators (ICD). ICDs have been used for many years in adults for a variety of problems but usually to treat life threatening heart rhythm problems, which may cause sudden death without any warning. The same things can happen to a child with severe hypertrophic cardiomyopathy (HCM). With the advent of more sophisticated ways to treat potentially life threatening heart rhythm problems, some parents are finding themselves presented with the option of having an implantable cardiac defibrillator (ICD) placed in their child.

ICDs have been used in children as young as 3 months old for the treatment of life threatening arrhythmias. They can be used to treat ventricular fibrillation (VF), preventing complications and possibly death caused from this serious life threatening heart rhythm. VF is a condition in which the heart’s bottom chambers, the ventricles, contract in rapid and unsynchronized rhythms. During this time, the heart cannot pump blood into the body, and as a result oxygen cannot reach the heart muscle, brain, and lungs. ICDs have been shown in some studies to prevent serious problems and/or death 90% of the time that they have worked (Heart Rhythm Society, 2006).

Some children with HCM are at greater risk of developing VF as a result of changes that have occurred in the heart muscle during the course of the disease. Your doctor can talk to you in detail about whether your child is at risk of developing VF and whether an ICD may be a treatment option.

An ICD is a small electronic device about the same size as a pager. It is similar to a pacemaker but works differently. Usually, the ICD is surgically placed through a small 3-inch incision just underneath the collarbone. Small cables (leads) connected to the ICD are threaded through a large vein directly to the heart’s lower chamber. The lead is then placed permanently in position in the right ventricle. Usually this surgery only requires an overnight stay in the hospital. Although this is not open-heart surgery, most children will need general anesthesia for the procedure.

ICDs are powered by small batteries. These batteries operate a tiny computer, which allows the ICD to work in the way it is programmed. Your child’s doctor can tell the ICD exactly when it should treat your child based on the heart rhythm that it detects. Fortunately, many children with HCM who have ICDs never have the need to receive a shock. However, if the need is there, it is important to have the ICD to treat the abnormal and potentially life threatening heart rhythm.

Children with ICDs lead normal lives and their restrictions are based on how serious their HCM is and not on the fact that they have an ICD in place. The ICD will likely not change any restrictions that your child’s doctor has placed on their activities.

FREQUENTLY ASKED QUESTIONS

Because many parents have never heard of ICDs or if they have, are not aware that children can have them, they may have many questions. Below are some common questions that parents and children have asked me before getting an ICD:

• Is an ICD the same thing as a pacemaker?

No. A pacemaker helps the heart’s own electrical system work when it has been damaged and can not produce normal heart rhythms. Usually a child with HCM will need an ICD to help treat and correct serious heart rhythms when they develop. A child’s natural pacemaker works fine. However, all ICDs do have a pacemaker inside of the device as well. This is used when your child’s own heart’s pacemaker doesn’t work quite right after a shock for VF. In this case, the ICDs pacemaker will help pace your child’s heart until his/her own heart’s pacemaker is working better.

• Can my child be around microwave ovens, computers, and cell phones?

Yes to all of them. None of these or other common household appliances alter the way an ICD functions. All newer cell phones are compatible with ICDs. We do ask that patients carry cell phones in shirt pockets on the opposite side of their ICD and that they listen and talk on them on the opposite ear. In general that is the only restriction.

• If I am near my child when he gets a shock, will I get shocked also?

No, the ICD only shocks the heart muscle and there is no conduction of electricity to others around him.
Can my child do everything he did before the ICD was implanted?
Your doctor can answer for your particular child, but in general, the answer is yes. Usually there is a period of weeks where there are some restrictions in arm movements and the weight that can be picked up with the arm on the side of the ICD. After the healing has taken place, your child should be able to live much like before the procedure.

Does the ICD have to get changed or does it last forever?
The ICD has a battery in it that will last for a period of years. The amount of time it lasts depends on a number of things. One of the most important factors in the battery life is how much your child uses the ICD. If your child uses the ICD as a pacemaker in daily life, the battery won’t last as long. In general, ICD batteries should last at least 4-5 years and maybe much longer. If your child is a heart transplant candidate, the ICD will be removed at the time of transplant.

How often do we need to go to the doctor to have the ICD checked?
Each doctor is different, but in general they like to check the ICD in the office every 3-6 months. The ICD is checked through a laptop programmer that is made specifically by the manufacturer of your child’s ICD. A small circular programmer head is placed over the ICD. The programmer reads what the settings are on the ICD and if any shocks have been given since the child was last seen. This is called an interrogation. The doctor or nurse doing the interrogation can tell if the ICD is working well. If your child has gotten shocked, they can look at what caused the shock and how the ICD responded to the abnormal heart rhythm. Additionally some ICDs can be checked over the telephone.

Studies have shown that children with ICDs adjust extraordinarily well to their devices and for the most part live very normal lives given their underlying heart problems. ICDs can help prevent serious complications of HCM and be used as part of an overall treatment plan for children with certain forms of cardiomyopathy.

April Perry works as a pediatric electrophysiology advanced practice nurse in the Duke Children’s Heart Program. She works with children and young adults who have cardiomyopathy and heart rhythm problems. April may be reached at 919-681-6772 or via email at perry042@mc.duke.edu.

Additional Resources on Implantable Defibrillators
Heart Rhythm Society
www.hrspatients.org/patients/treatments/cardiac_defibrillators/default.asp

American Family Physician - The Implantable Cardioverter-Defibrillator.
www.aafp.org/afp/980115ap/980115b.html

Heart Center Online for Patients - The ICD Center
www.heartcenteronline.com/The_ICD_Center.html

Websites of Interest
Two websites of interest to both families and physicians are the National Society of Genetic Counselors (www.nsgc.org) and the Congenital Heart Information Network (www.tchin.org). The NSGC is a professional society that promotes the genetic counseling profession as an integral part of health care delivery, education, research, and public policy. CHIN is an international organization that provides reliable information, support services, and resources to families of children with congenital heart defects and acquired heart disease.

Features offered on the NSGC website include searches for genetic counselors by zip code, by name, by institution, and by area of practice or specialization. With genetic discrimination such an important topic these days, NSGC has links to sites that contain genetic information laws, position statements, advocacy resources, and reports on discrimination. It also offers an extensive booklist with descriptions and links to reviews. There is also a list of the most common questions that are asked about genetic counseling, and instructions on drawing a family tree so that families can keep track of their medical history and possible health risks of previous generations.

The CHIN website offers several resources of interest to patients and families including online discussion groups, family galleries, CarePages, book reviews, and listings of local support groups. The Resource Room section presents informational materials written and reviewed by health professionals. The site also provides links to other websites such as those of pediatric cardiology departments and congenital heart surgery programs throughout the world.
On Thursday September 14, 2006, the Children’s Cardiomyopathy Foundation hosted its Fourth Annual Golf Classic at the New York Country Club in New Hempstead, New York. Despite the inclement weather, a sizable group of devoted supporters came out and raised over $270,000 gross, surpassing last year’s total by 24%. As in years past, the event was heavily supported by the financial services industry with more than 30 investment and commercial banks, hedge funds, and law and money management firms participating. This year showed a marked increase in corporate sponsors due to the hard work of the event committee, chaired by Ian Sandler of Morgan Stanley.

The Annual Golf Classic is CCF’s main fundraising effort, and net proceeds from this year’s event will be earmarked for CCF’s research grant program. Each year, CCF awards research grants to promising investigators to support basic, clinical, or translational research related to the cause or treatment of cardiomyopathy in children. Since 2002, CCF has committed over $600,000 towards pediatric cardiomyopathy research and treatment initiatives.
New Jersey Appraisal Institute Supports CCF

The Metro New Jersey chapter of the Appraisal Institute hosted a golf tournament on June 22 at the New Jersey National Club to benefit CCF. The outing raised $5,500 for CCF’s research and education fund. The Appraisal Institute is an international organization that supports education for real estate appraisers throughout the world. President Denise Smith chose CCF as the beneficiary of their annual event when board member Victor DiSanto nominated CCF. Victor’s son, Joey, 12 was diagnosed with dilated cardiomyopathy a year ago.

Joey, a versatile athlete, started playing golf with his father when he was diagnosed with cardiomyopathy. For the sake of his health doctors advised him that golf was the only sport he could safely engage in. Although he was scheduled to play, on the day of the event Joey was in the hospital waiting for a heart transplant. “We never anticipated that the person we were doing this for would be in the hospital at the time,” says Debra Miller, Executive Director of the Appraisal Institute. Fortunately, Joey received a donor heart on July 8 and is now doing well post transplant.

Mom Inspired T-shirt Raises Awareness

CCF family member, Jennifer Dickson, was training to run a 5 person relay marathon when she had the idea to run the race in honor of her 2 1/2 year old son, Drew, who has HCM. Jen wanted to create a simple t-shirt that would bear the names of other CCF children affected by cardiomyopathy so she could run the race in honor of them as well. Since the idea was a big hit with CCF families and friends, she decided to produce the shirts in bulk and sell the shirts to raise money for CCF. Nearly 290 t-shirts were sold, raising over $2,600 in net proceeds.

CCF still has a limited number of t-shirts (large size only) available for $16 each. To place an order, please contact Harriet Salk, at hsalk@childrenscardiomyopathy.org. CCF also has other t-shirt designs available from our online merchandise shop, www.cafepress.com/cardioxmlpathy.

2nd Annual Neiman Marcus Event in Michigan

On April 30, CCF partnered with Neiman Marcus, the high-end specialty retailer, for its third fundraiser in Troy, Michigan. CCF family member, Jennifer Ghandour, whose sons Anthony, 7, and Eli, 5, have hypertrophic cardiomyopathy, served as the event chairperson. Nearly 50 people attended the “Swing into Spring” champagne brunch and fashion show, raising close to $3,500. The event also included a raffle of items such as jewelry, spa certificates, hotel stays, and weekend get-aways. Local merchants donated nearly $5,000 worth of goods for the raffle.

CCF families from the Michigan area were able to meet each other for the first time. “It was nice because all the families from around here who are affected by cardiomyopathy were able to get together,” says Jennifer. CCF family members Scott and Penni Newport and their son Evan attended, as did CCF staff member Stormy Hill and her family.

Brian and Angela Hill, whose daughter Andri had restrictive cardiomyopathy but received a heart transplant in 2005, spoke at the event. Other speakers included Jennifer herself and Dr. Richard Humes, Chief of Pediatric Cardiology at the Children’s Hospital of Michigan. “It was a lovely event, and I’m glad I was able to help CCF. I look forward to the next one,” Jennifer added.

(L to R) Scott Newport, Jennifer & Anthony Ghandour, Stormy Hill, Fayette Waligorksi, Angela & Brian Hill

Annual Salem Road Race Raises Funds for CCF

Wearing bright-red t-shirts sporting the CCF logo, over 165 runners made their way across the finish line of the 13th Annual Salem Road Race on March 18 in Salem, Connecticut. Thanks to the efforts of Rick and Gina Konon, whose three-year-old nephew has hypertrophic cardiomyopathy, the 5K and 1-mile family-fun run raised close to $2,100 for CCF and increased public awareness about pediatric cardiomyopathy.

The popular community race event was at risk of being cancelled this year. So Rick, an avid runner; and his wife Gina took it upon themselves to organize the race and to raise funds for CCF at the same time. They saw this opportunity as the perfect way to support CCF’s mission. They went into action obtaining sponsors, in-kind donations, insurance, awards, and t-shirts. “For weeks, I worked at home instead of the office so that I could dedicate as much time as possible to organizing the race. I love running and of course, we love Aidan our nephew,” says Rick.

The Konon family of six was out in full force on race day, with several members running in the race and assisting with the overall management of the event. With the race securely back on the road racing calendar for 2007, Rick and Gina hope to raise even more money this coming March on behalf of CCF and Aidan.
New Research Study on HCM

There is a new pilot study being done to investigate whether the drug, Diltiazem, a calcium channel blocker, can attenuate the natural history of disease development of hypertrophic cardiomyopathy (HCM). Diltiazem is FDA approved to treat hypertension and heart rhythm disturbances. It has a long track record for safety, but its use for HCM is experimental. This study was initiated after promising results in the mouse model of HCM performed in the laboratory of Jon and Christine Seidman at Harvard Medical School/Brigham and Women’s Hospital.

Dr. Carolyn Ho, Medical Director of the Hypertrophic Cardiomyopathy Clinic at Brigham’s Women’s Hospital, is the principal investigator of this single center study. She is working with Dr. Steve Colan at Children’s Hospital Boston to include children (greater than 5 years of age) in this clinical trial.

Current management strategies for HCM focus on treating symptoms. There are no available therapies that address disease prevention. This study would be one of the first to look at a strategy of early treatment to minimize or interrupt the development of the disease. Most likely, this pilot study will be expanded in scope to include multiple centers in the future.

Patient Enrollment Criteria
Eligible subjects will have a known sarcomere gene defect associated with HCM but with no clinical symptoms of cardiomyopathy and no indication of left ventricular hypertrophy on echocardiography or EKG. Children age 13 and older will be enrolled at Brigham and Women’s Hospital; children age 5-12 years will be enrolled via Children’s Hospital Boston.

Contact Information
For more information or to enroll in this study, please contact Allison Cirino at 617-732-7921 or acirino@partners.org. Please refer to this study by the www.clinicaltrials.gov identifier NCT00319982. Dr. Carolyn Y Ho can also be contacted at 617-732-7367 or cho@partners.org

Annual Appeal Campaign Underway
A Little Heart Can Go a Long Way...

This year, remember that a gift from the heart can make all the difference...

Please consider giving to CCF’s annual appeal. With your support, we can continue to advance research and education on pediatric cardiomyopathy, giving affected children and their families hope for a brighter future.

A donation and company matching gift can be made using the enclosed remittance envelope.

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