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Living with an ICD… A Story of a Family, a Disease and an Industry Nearly 60 Years in the Making

By Lisa Salberg, CEO and Founder
Hypertrophic Cardiomyopathy Association

How does one explain what it feels like both physically and emotionally to live with cutting-edge technology implanted directly into your heart? To answer this question you have to go back and understand why a person would find themselves in the position of needing to use this technology. Every person's story is different, every family's story is different. This one is my story, and we have to go back to 1953 to understand it completely.

It was June in northern New Jersey, and my father, a 17-year-old boy, took his girlfriend out on a date. With graduation from high school the next day, life was looking good -- until he got home. His mother was frantic about something. It was his father, his 43-year-old strong healthy carpenter father was not well; he was not breathing. While yes, they did tell him to stay away from caffeine because it made his heart pound, and there was some question whether or not his heart sounds were completely normal, but many people have heart murmurs, right? Until this night, this was a healthy 43-year-old man. Larry Flanigan Sr., my grandfather, died that night. Our family would never be the same, and the wounds would take decades to heal, if in fact they ever really did.

Flash forward to the early 1960s my great aunt, had some heart problems. "She died unexpectedly," I was told. She had a stroke in her mid-50's. A few years go by; it is now the late 1960s - early 1970s, and my uncle was told he had idiopathic hypertrophic sub aortic stenosis IHSS. Shortly thereafter, my sister received the same diagnosis. Several years later it was my turn; in 1979 I was diagnosed with IHSS. While we all took it in stride, and for the most part, we didn't let it dictate who we were or how we lived our lives. In some ways this was a good thing; in other ways it may have lead to some leaving way too soon.

My uncle packed up, left New Jersey, and moved to Montana. He lived in a very beautiful, but remote area. While he enjoyed outdoor activities, including hunting, hiking and even panning for gold in the Rocky Mountains, his heart could not take it. It was August 1990 when we got the call telling us that he was found dead in his car outside a sportsman show in Helena, Montana; he was 47 years old.

What I failed to mention was that his death came within hours of me having a Hickman catheter removed from my chest that was finishing my treatment for subacute bacterial endocarditis, which seven weeks earlier had caused me to have a stroke at the ripe old age of 21. While I would like to say that I have always been the best patient - always seriously taking care of myself, aware of my disease and always embracing a healthy relationship with the healthcare community, it would not be true. I blindly trusted others to tell me what to do, and no dentist ever told me I needed antibiotics

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before dental work. I learned a critical lesson: it only cost me a portion of my brain and eyesight. While I always wanted to think my 'heart problem' was a minor and manageable thing, recent events made me stop and reevaluate the reality of the situation. There was no fighting this any longer; it was time that our family took this disease very seriously; it was time we all got more aggressive.

We were involved in a research program; I wish I could say it was a good experience; it was not. While it does not seem that long ago, research for an uncommon disease in the late 1980s, early 1990s was very different from where we are today. My family was manipulated, facts and treatment options withheld from us, and we were strong-armed into consenting to tests and procedures, and devices which we later found had no scientific or therapeutic value. It was during this time that my sister Lori received her first pacemaker, supposedly to protect her and to somehow return her heart to a more normal function. About a year later, I received my first pacemaker; it was now 1992. The scientific literature played out the story of DDD pacing in HCM in very clinical terms. What is always lacking from these documents are the human implications and emotional costs.

Lori's health spiraled out of control as I watched her move from medication to medication, and then to changing pacemaker settings. Everything just seemed to slip through the cracks, and nobody took her complaints seriously. The hometown physicians would point to research specialists and vice versa; she was caught up in an awful spiral. While this was all happening, each step seemed to make sense, yet now when you look back at each one, something seems wrong. It was now 1995. Admittedly, I was not a hundred percent focused on my sister's health at that time, as I was pregnant with my daughter and attempting to manage having HCM, a full-time job, a home and the pregnancy.

June 11th 1995; 9:30 p.m., Sunday - it is amazing how some dates and times can stick in your mind. I can tell you exactly what I was doing: I was on the phone with my sister Lori, thanking her for the baby shower she had thrown for me a week earlier, talking about my niece and nephew, and asking for my big sister's guidance as to what to expect when I was delivering my daughter. It was so normal. We ended the call the way we always did… wrapped it up, and said, "I'll talk to ya later – bye bye." I had no idea that I would never talk to my sister again. She went to bed shortly after she hung up the phone with me. My phone rang at 5:55 a.m., June 12, 1995; it was my mother telling me my sister was not breathing and paramedics were en route. Lori had suffered sudden cardiac arrest in her sleep. The next five days were a roller coaster of emotion for a thousand reasons as you can well imagine. It was Wednesday, June 14 when the EEG was done, and there appeared to be no brain activity; it appeared my sister was gone. We wanted to give her 48 hours, time enough for a miracle, and enough time for her children and the rest of us say what we needed to say, do what we needed to do.

June 16, 1995, 4:36 p.m. I never recalled seeing the cardiologist cry before - my sister's hometown cardiologist arrived to check "one last time," and the reality of the moment did not feel quite real to me. Here I was, her eight-month pregnant sister watching all this, knowing that I had the same heart condition. The doctor did not come to offer care or treatment; he came to pronounce her dead. He checked her vitals, looked at her chart, held her hand, kissed her forehead and shed a tear. I could tell you that I was at peace with this, but that would be a huge lie. My 36-year-old sister was dead, and there were simply no feelings to describe this. My daughter would never know her Aunt Lori, and to this day that thought makes me cry, her children were, for all intent, parentless, my parents had to bury a child, and I lost my best friend.

A month later my daughter was born – and I crashed in delivery (a drama for a different talk!). From the moment I saw her, I wondered did she have HCM too? It would take seven years to know. I found myself in a place I never expected to be - raising my sister's two children, a baby of my own, and all with a heart that seemed, based on history, not to be built for longevity. The only thing I could think of was "I'm too damn busy to die." I have to protect myself. I sought information from the leading experts in the field, I extricated myself from the institution that led my sister so far astray, and I began to form the *Hypertrophic Cardiomyopathy Association* (HCMA). I turned my inquisitive nature toward understanding the enemy within, and arming myself with every tool in the arsenal to ensure that I had a chance to finish the work that I must, and to ensure my children have a better future than what history has handed them.

In March of 1996, I was told I needed an Implantable Cardiac Defibrillator (ICD). Given my relatively good health, lack of previous cardiac arrest and apprehension for the healthcare community, I weighed this choice very carefully and became a student of the device industry. Be it right or wrong, my choice in 1996 was to wait until technology caught up with me a little more. I had a pacemaker which really wasn't doing much for me at the time, but I was concerned about not being paced any more; it was uncharted territory. The first thought was to implant an ICD on one side of my chest, pacemaker on the other with two sets of wires. I am no engineer… but the thought of all those wires and all that electricity in my body made me think… there is too much room for error. One manufacturer was in trials for dual chamber piece or defibrillator unit and another manufacturer was hot on their trail. I opted to wait for one of these devices to come to market to get my device, about an 18 month window. In July 1997 the first dual chamber piece or defibrillator was approved in the United States by the FDA. Within 24 hours of approval I received a phone call from the out-of-state physician who I had made arrangements with to do the implant once it was available. Within five days I was in the hospital having my pacemaker taken out and my ICD implanted. That device lasted until 2001. I experienced a device failure as a result of the solder break within the unit. I had no adverse effects from the failure. On April 2, 2001, I had my second ICD implanted, and it lasted a short period of time. The battery depleted much quicker than we had anticipated. As of 2010, I am on my fourth implantable device.

My ICD was indicated for several reasons including the underlying diagnosis of hypertrophic cardiomyopathy. My personal risk factors were: a family history of sudden cardiac arrest, hypotensive blood pressure, a history of ventricular arrhythmias and recurring syncope. A common question I am often asked is "has it ever gone off?" Nearly 13 years with an implantable defibrillator and not so much as one shock, yet tons of reassurance and peace of mind.

It took until the early 1990s to figure out that yes, my father also had HCM. Dad received his first device (bi-ventricular pacer/ICD) in 2001, just days after 9/11 in a hospital overlooking the destruction of the World Trade Center. In 2002 my 19-year-old niece had an, ICD implanted; to-date her devices have not gone off; She is stable and well at the age of 25.

The trickier situation came in 2005. We learned through genetic testing in 2002 that my daughter carried genetic mutation for HCM; she was seven at the time. Three years went by with no problems and the symptoms and all echoes were clear, then she started complaining about her heart racing. She would wake in the middle of the night yelling "Mommy my heart," her pulse would be 160 – 170, BP always low. After a few events like this we had an echocardiogram performed which found her to be mildly abnormal. After a cardiac MRI, it was clear she was her mother's daughter, her heart looked very much like mine. The choice to implant an ICD in your own child is much harder than the decision to implant one in yourself. The implications for her lifetime, this social stigma, the impact on an adolescent girl's self-esteem, the worrying (What if I did not implant it and she needed it – could I live with myself?) all caused me many sleepless nights. In the end, I opted to talk to my daughter, and let her be part of the decision-making process to have or not have an ICD implanted in her chest. Sometimes the simple wisdom of a child makes things clear. She said, "Mom you have one, Stacey has one, and Grandpa has one, and I think I would feel safer with one too." She chose the device. On March 2005 my 10-year-old daughter received her first ICD. Three uneventful years went by. My daughter loves to ride horses and has since the age of eight. On a September afternoon, 'a horse had a bad day,' and unfortunately, my daughter was on that horse. While she controlled the horse as it was in a full gallop, it was terrifying experience for her, and her heart rate went way above the threshold of the device. She received two heart shocks while on the horse. The third one caused her to jump to the ground. Many questions plague my mind about that day. If she was riding a horse and did not have a device, would that event have led to fatal arrhythmia? if she was riding a horse, and did not have the device, would she have gotten off the horse without incident? I don't know, nor does anyone else.

So how do I answer the question, "How does it feel to have life-saving technology implanted into your chest?"

I embrace technology, I am grateful for technology. I can only hope that it not only continues to improve, and that all those that need an ICD have access to utilize it. Knowing you have an emergency room in your chest 24 hours a day, 7 days a week brings great comfort and freedom.

I would say that I have a rather unique point-of-view when looking at some of the other issues facing those with devices. When the HCMA was founded in 1996, only a handful of us had ICD's. In fact, few people in the world had these devices. Today there are millions of people worldwide with this technology keeping watch on their hearts. As of 2010, I have had the honor of working with over 4,700 families worldwide with a diagnosis of HCM. Listening to many of their concerns, pre- and post- implant has been enlightening. We have learned a great deal about HCM in the past decade, including that one risk factor for sudden death is ample to consider ICD implantation. In the past 3 weeks alone, I have spoken to 3 families who learned that lesson the hard way… they were waiting for more risks before the device was implanted. Let's take a quick look at them:

- A 44-year-old man who died in front of his 2 kids (only risk factor was family history).
- A 10-year-old boy who died in his parent's arms at home (only risk factor massive hypertrophy).
- A 51-year-old woman who is now severely brain damaged after a SCA, save (only risk factor, recurrent syncope).

The evolution of the device industry has not always been a smooth path, and the industry, physicians and patients have had to weather some storms together. While I could write a complete book on this topic alone, I will subject my comments here to several bullet points and focus on the patient side of where we can make improvements.

Patients do not always fully appreciate the implications of living with this technology, and what affects the device may have on them years down the line. When a patient hears, "you are at risk for sudden cardiac arrest, and you need an implantable cardioverter defibrillator…"

- It is rarely a pleasant message to receive…
- It often caused intense feelings ranging from fear to resignation…
- It is rarely an easy concept to absorb…
- Some patients consent to the device before fully understanding the entire situation…
- Few patients really fully understand the technology (heck many of us don't know how MP3 players work either!)
- Some people think that their device will protect them from all heart problems… this is not accurate…
- Few patients think of THEIR role in device safety and reliability…
- Few patients really think about the reality of an advisory or a recall of their device… but it is likely to happen to many of us…
- Few of us think about the day when we say… enough… it is time for me to let nature take its course.

Areas that patients are not thinking of at the time of implant, most specially include device alerts or recalls. The reality is that devices will be recalled, reliability will change, and we may face information about our device from seemingly out-of-the-blue.

This does not mean you just won the "recall lottery" – be wary of ads from law firms and offers of big pay outs.

Devices are manmade devices and subject to failures. A patient should understand that their role in keeping up on interrogations of the device will help to ensure they are notified in a timely manner of any alerts, recalls or other important data about the device.

Until 2005-6 manufacturers did not view patients as customers, they saw the doctor and the hospital as their customer.

A few of us spoke out… and demanded they communicate directly with us. Now device manufactures communicate with physicians first, then patients or consumers directly via a letter.

Good news… they communicate with us… bad news… the supreme court stripped us of many rights to recourse in the courts in the event of a device failure….3 years later.

Now can I say that I think that we as patients have a strong enough voice in matters of importance to the device industry? I am sorry to say the answer is "no." While we know devices are strong, reliable and backed by great science, we also know that they are manmade devices and thereby, subject to failures.

The Medical Device Act is an important piece of legislation that patients should be aware of. It is needed due to an over-reaching by the Supreme Court in Riegel v. Medtronic. The Supreme Court made it impossible for a consumer of an implantable medical device

to seek recourse in state courts if they are harmed by a device. This creates a very interesting situation, and one that I can find no other like in the United States. It appears that implantable medical devices are the ONLY consumer product that the consumer has no way to seek damages, if in fact, they are harmed. This creates an air of concern, and should be corrected with the passage of the Medical Device Act. There is some chance that this language will be folded into health care reform, but that has yet to be worked out, along with a long list of other important issues.

Lastly, it is important for patients to not only understand their devices, that they have a responsibility to make sure if they move to notify device manufacturers, and to have their devices interrogated regularly… but to know WHY they have a device to begin with. I am constantly amazed when attending functions for those with devices that many have no idea what their underlying diagnosis is. Many reasons for devices are genetic in nature and knowing why a family member has a device, may have a direct impact on other family members.

In closing, you can live a long, active and productive life with an ICD and thousands of people do. Ensuring that patients and their families understand the device and the reason the device is being implanted is critical to ensuring that the patient's life will be positively impacted by the device. I travel a great deal, live a 'normal' life, and know I am protected against SCA. I am very grateful for my, my daughter's, my niece's, my cousin's and all of my friends' ICDs each and every day. In a perfect world all those at risk for SCA would be aware of their risks, and be offered the option of a device. In the end, to receive a device is a personal choice. It is a choice one should make with all available data about the device, knowledge of the expected outcome of the disease and with the assurance that we are entering into a partnership that sees us, the patient, as a partner in our futures together with the device industry, the health care community and our families.

CCT

This article is based on a speech given by Lisa Salberg, entitled, "Living with an ICD - From a a Patient's Perspective" at the University of Colorado ICD Support Group on February 2, 2010. Dr. Michelle Khoo MD, Cathy Kenny, NP and Rick McLaughlin, BBA were available before and after the presentation for questions.

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SEPTEMBER
MEDICAL MEETING FOCUS

Pediatric & Congenital Rhythm Congress
September 22-25, 2010; Çeşme-Izmir Turkey
www.pedirhythm.org

Congress Directors: Alpay Çeliker, George F. Van Hare, Richard Friedman, Joachim Hebe

Overview: This 2-day congress will cover: fetal and pediatric clinical arrhythmia recognition and management, 3-dimensional mapping, ablation therapy, and device therapy. A particular focus will be on the abnormal repolarisation syndromes. Keynote lectures will be delivered by a distinguished international faculty, while a large number of selected oral communications and poster presentations will report new data about the most sophisticated methods from around the world.

The Congress will feature panels, case discussions, debates, and lectures, offering attendees the chance to be updated on recent innovations in technology, and to learn the most recent state-o- the-art in the diagnosis and management of arrhythmias, and in innovative technology.

Program includes:
- Postoperative Dysrhythmias
- Case Discussions
- How To?
- Fetal Arryhythmias
  - Diagnosis and Evaluation
  - Treatment
- Ablation:
  - Hot Topics
  - Atrioventricular Nodal Reentrant Tachycardia
  - Wollf-Parkinson-White Syndrome
  - Postoperative Hearts/VT
- Pacemaker & Defribulators:
  - Basic Knowledge
  - Pacemaker Follow-up
  - Hot Topics
  - Cardiac Resynchronization
  - Defibrillator Follow-up
- and more...

Invited Faculty: Kamil Adalet, Özgür Aslan, Ahmet Akyol, Kudret Aytemir, Charles Berul, Nico Blom, Konrad Brockmeier, Alpay Çeliker, J. De Giovanni, Erdem Diker, Aygün Dindar, Fabrizio Drago, Okan Erdoğan, Cengiz Ermiş, R.A. Friedman, Helena Gardiner, Bülent Görenek, Can Hasdemir, Joachim Hebe, Gabrielle Hessling, Jan Janousek, Peter Karpawich, Ayhan Kılıç, Ata Kırılmaz, J. Kirsh, Sedat Köse, Osman Küçükosmanoğlu, Jerome Le Bidois, Enis Oğuz, A. Oto, Bülent Özin, Sema Özer, John Papagiannis, Thomas Paul, Andreas Pflaumer, E Rosenthal, Arda Saygılı, Michael Schaffer, Klaus Schmidt, Massimo Silvetti, İzzet Tandoğan, John K. Triedman, George F. Van Hare, Artur Wilde, and Christopher Wren

Visas are required; contact your nearest Turkish Embassy or Consulate to check for the visa regulations.

Abstracts from "Evolving Concepts in the Management of Complex Congenital Heart Disease II" - Part II

"Abstracts from 'Evolving Concepts in the Management of Complex Congenital Heart Disease II' - Part III" includes the following topics and presenters:

- Clinical Databases: Status and Utility *by Jeffrey P. Jacobs, MD, FACS, FACC, FCCP*
- Sudden Cardiac Death and Channelopathies *by Joel A. Kirsh, MD*
- Diagnosis and Management of Cardiomyopathy in Children by *Steven Lipshultz, MD*
- A Program of Fetal Intervention for Hypoplastic Left Heart Syndrome: Lessons Learned in the First 10 Years *by Audrey C. Marshall, MD*
- 3-D Echocardiography Current Status and Uses *by Gerald R. Marx MD*
- Surgical Strategies to Maximize Neurological Outcomes *by Peter Pastuszko, MD*

Read Parts I and II in the April and May issues of Congenital Cardiology Today.

Abstract Title: Clinical Databases: Status and Utility

Presenter: Jeffrey P. Jacobs, MD, FACS, FACC, FCCP; Surgical Director of Heart Transplantation and ECMO; The Congenital Heart Institute of Florida (CHIF); St. Petersburg, FL USA

Objective

In order to perform meaningful multi-institutional analyses, any database must incorporate the following six essential elements:

1. Use of a common language and nomenclature,
2. Use of a database with an established uniform core dataset for collection of information,
3. Incorporation of a mechanism to evaluate case complexity,
4. Availability of a mechanism to assure and verify the completeness and accuracy of the data collected,
5. Collaboration between medical and surgical subspecialties, and
6. Standardization of protocols for life-long longitudinal follow-up.

The objective in this presentation is to review the current state-of-the-art of clinical databases focusing on the six areas listed above.

Abstract

During the 1990s, both The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons created databases to assess the outcomes of congenital cardiac surgery. Beginning in 1998, these two organizations collaborated to create the International Congenital Heart Surgery Nomenclature and Database Project. By 2000, a common nomenclature, along with a common core minimal dataset, were adopted by The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons, and published in the Annals of Thoracic Surgery. In 2000, The International Nomenclature Committee for Pediatric and Congenital Heart Disease was established. This committee eventually evolved into the International Society for Nomenclature of Paediatric and Congenital Heart Disease. The working component of this international nomenclature society has been The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease, also known as the Nomenclature Working Group. By 2005, the Nomenclature Working Group cross-mapped the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons with the European Paediatric Cardiac Code of the Association for European Paediatric Cardiology, and therefore created the International Paediatric and Congenital Cardiac Code, which is available for free download from the internet at www.IPCCC.NET.

This common nomenclature, the International Paediatric and Congenital Cardiac Code, and the common minimum database data set created by the International Congenital Heart Surgery Nomenclature and Database Project, are now utilized by both The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons. Between 1998 and 2009 inclusive, this nomenclature and database was used by both of these two organizations to analyze outcomes of over 200,000 operations involving patients undergoing surgical treatment for congenital cardiac disease.

Two major multi-institutional efforts that have attempted to measure the complexity of congenital heart surgery are the Risk Adjustment in Congenital Heart Surgery-1 system, and the Aristotle Complexity Score. Current efforts to unify the Risk Adjustment in Congenital Heart Surgery-1 system and the Aristotle Complexity Score are in their early stages, but encouraging.

Collaborative efforts involving The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons are under way to develop mechanisms to verify the completeness and accuracy of the data in the databases. Under the leadership of The MultiSocietal Database Committee for Pediatric and Congenital Heart Disease, further collaborative efforts are ongoing between congenital and paediatric cardiac surgeons and other subspecialties, including paediatric cardiac anesthesiologists, via The Congenital Cardiac Anesthesia Society, paediatric cardiac intensivists, via The Pediatric Cardiac Intensive Care Society, and paediatric cardiologists, via the Joint Council on Congenital Heart Disease and The Association for European Paediatric Cardiology.

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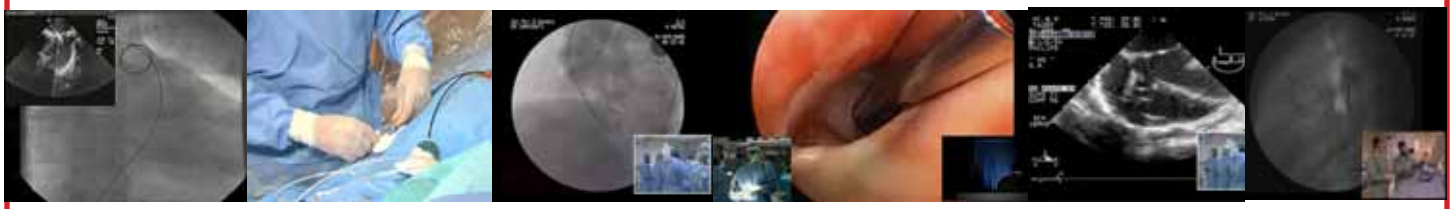

- Percutaneous Pulmonary Valve Implantation
- Pulmonary Artery Rehibilation
- Device Closure is Safe & Should be Continued
- Catheter Management in the Neonate
- Duct Dependent Lesions: Stent vs. Shunt
- Intervention in the Immediate Post-op Period
- The Edwards Valve
- Percutaneous Closure of the VSD
- Transcatheter Implant of a Melody Valve Watch - VSD Closure
- Covered Stent to Eliminate Fontan Baffle Leak & Stenosis
- Native Coarctation of Aorta - Stent Implantation
- Pulmonary Artery Flow Restrictors
- Transcatheter Valve
- Intra-operative PA Stent
- Perventricular Muscular VSD Device Closure
- Closure of Septal Defect Using Real Time 3D Echo Guidance
- Perventricular Membranous VSD
- Hybrid Stage I Palliation for HLHS PA Bands and PSA Stent
- Intraoperative Aortic Stent for CoA
- Intraoperative LPA Stent Using Endoscopic Guidance
- Creation of ASD after PA Bands & PDA Stent for HLHS in a Preemie
- Perventricular Implant of Edwards Valve Stent in the Pulmonary Position
- Closure of Septal Defect Using Real Time 3D Echo Guidance
- High Frequency Ultrasound Creation of ASD
- PmVSD Closure
- Hybrid Stage I Palliation for Complex Single Ventricle in a 1.4 kg Neonate
- Transcatheter Implantation of Implantable Melody Valve
- Perimembranous VSD Closure with Amplatzer Membranous VSD Occluder
- Stent RPA, Pre-Stent Conduit & Melody Valve Implant
- Transeptal LHC Assessment of Residual Coarctation
- Coarctation Stenting Using a Covered CP Stent
- Closure of ASD Under ICE Guidance
- PFO Closure
- Transcatheter Closure of Atrial Septal Defect (ASD)
- Closure of ASD under TEE Guidance
- Transcatheter Closure of Moderate PDA
- Pulmonary Angioplasty
- Hybrid Stage Palliation
- ADOI or MVSD Closure of PDA
- VSD Closure Using a PDA Device
- Balloon Pulmonary Valvuloplasty
- Coil or Vascular Plug II closure of Coronary Fistula
- Device Closure of ASD
- Stent Placement Across Recurrent Coarctation
- Stenting RVOT on 7-day Old Newborn
- Transfemoral AVR for AS
- ASD Closure with Septal Occluder
- Stenting of Coarctation of Aorta
- Transapical AVR for Aorta Stenosis
- and more….

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Analysis of outcomes must move beyond mortality, and encompass longer term follow-up, including cardiac and non cardiac morbidities, and importantly, those morbidities impacting health related quality of life. Methodologies must be implemented in these databases to allow uniform, protocol driven, and meaningful, long term follow-up.

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CCT

Abstract Title: Sudden Cardiac Death and Channelopathies
Presenter: Joel A. Kirsh, MD; Labatt Family Heart Centre; Hospital for Sick Children; Toronto, ON, Canada

Objective

Attendees will acquire new knowledge regarding the prevalence and presentation of electrical heart disease(s) as the cause of sudden death, and understand the approach to investigating families for the presence of these diseases. Attendees will then be able to make important diagnoses in pre-symptomatic carriers of potentially lethal cardiac rhythm disorders, and intervene before tragedy strikes the family again.

Abstract

Although proportionally rare compared to the older adult population, sudden cardiac death (SCD) occurs in children and young adults, and due to the number of productive life-years lost, approaches a similar impact on society. More than half of SCD is due to structural or acquired heart disease that is usually evident on postmortem examination, but a significant number of these deaths remain unexplained. With increasing appreciation of the prevalence of cardiac channelopathies, and the growing availability of both clinical and molecular testing, the causes of previously unexplainable cases of SCD may now be determined, and family assessments may be conducted to identify patients with potentially lethal cardiac rhythm disorders before tragedy strikes. This presentation will review the current data and guidelines in the field as well as present the results of investigative strategies for the diagnosis of these disorders.

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CCT

Abstract Title: Diagnosis and Management of Cardiomyopathy in Children
Presenter: Steven Lipshultz, MD; Professor and Chairman of Pediatrics Chief of Staff of Holtz Children's Hospital; University of Miami; Miami, FL

Objective

1. To identify the importance of cardiomyopathy as a source of significant morbidity and mortality in children.
2. To review the current evidence that supports the best diagnostic and therapeutic approaches to these children to optimize clinical outcomes.
3. To demonstrate the need for more evidence-based studies to identify patient-specific therapies to positively impact the outcomes in this population of children.

Abstract

Cardiomyopathy is a serious disorder of the heart muscle, although accounting for only 1% of pediatric cardiac disease, it results in a disproportionately greater significant morbidity and mortality. Nearly 40% of symptomatic children die within two years after diagnosis unless they receive a heart transplant and a large proportion of those who survive without a heart transplant are left with permanent myocardial damage and dysfunction. As a rare disease, diagnosis and management is not guided by robust clinical trials. Observational studies provide the best evidence for the treatment of these patients at this time.

The Pediatric Cardiomyopathy Registry (PCMR), funded by the NHLBI since 1995, was designed to describe the epidemiology and clinical course of selected cardiomyopathies in patients 18 years old or younger and to promote the development of etiology-specific prevention and treatment strategies. As of December 2009 the PCMR

has collected data on 3,549 patients across 98 clinical centers with nearly 40,000 completed forms and more than 14,000 patient-years of follow-up.

Using PCMR data, the incidence of cardiomyopathy in two large regions of the United States is estimated to be 1.13 cases per 100,000 children. Only 1/3 of children had a known etiology at the time of cardiomyopathy diagnosis. Using PCMR data, a diagnostic algorithm has been developed which is intended to increase the determination of specific etiologies and decrease the number of children categorized as having idiopathic disease. Numerous PCMR studies have identified the risk factors associated with poor outcomes in primary or idiopathic dilated, hypertrophic, restrictive or left ventricular non-compaction cardiomyopathies, or those as a result of myocarditis or genetic syndromes.

Secondary cardiomyopathies can arise from exposure to HIV or anthracycline therapy for pediatric cancer. Results from both observational and clinical trials, supported by both the NHLBI and NCI, have identified risk factors for negative outcomes in these patient groups, as well as treatments than can ameliorate risk in these populations.

Analyses of PCMR data regarding standard of care and real-world medical therapy offered to children with DCM shows that treatment has not significantly changed over the past few decades. This finding illustrates the need for further pediatric-specific investigations in order to define evidence-based therapies. However, the very few clinical trials conducted in this population have found no evidence-based effective therapies. Failing conventional management, the only treatment option for patients is heart transplantation. Currently, the PCMR and Pediatric Heart Transplant Study databases have been merged resulting in the identification of factors associated with adverse outcomes in patients awaiting or status post heart transplantation. These results will lead to better informed clinical decisions about patients, most likely to benefit from heart transplantation.

In rare diseases like pediatric cardiomyopathy, clinical decisions regarding screening, diagnosis, and treatment are usually based on observational studies. We present the results of such studies here to assist clinicians in decision-making in this population which experiences high morbidity and mortality. The results of these studies can lead to better diagnostic strategies, improve patient outcomes, improve clinicians' prognostic abilities, potentially improve the selection of patients most likely to benefit from heart transplantation, and clearly demonstrate the need for further development of evidence-based best practice measures.

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CCT

Abstract Title: A Program of Fetal Intervention for Hypoplastic Left Heart Syndrome: Lessons Learned in the First 10 Years
Presenter: Audrey C. Marshall, MD; Chief, Invasive Cardiology; Children's Hospital Boston; Boston, MA USA

Objective

The attendees will be familiar with the rationale for, and techniques of, fetal aortic valve dilation for prevention of Hypoplastic Left Heart Syndrome. They will gain the competence to provide fact-based preliminary counseling to potential candidates. Armed with the current indications, requirements of candidacy, and procedural outcomes, the attendees will gain the ability to improve overall outcomes of fetal aortic stenosis by identifying fetuses who may achieve biventricular repair in the neonatal period.

Abstract

Neonatal HLHS encompasses a spectrum of left heart lesions, some of which are identified prenatally as aortic stenosis with a normal-sized or dilated LV. Associated features in those who progress to HLHS include retrograde flow in the transverse arch, left-to-right flow at the foramen ovale, and monophasic mitral valve inflow. This constellation marks those fetuses with progressive left heart hypoplasia. Since 2000, we have pursued a program of aortic valve dilation in mid-gestation fetuses with AS in an effort to interrupt the natural history of HLHS. We hypothesized that relieving LV outflow tract obstruction would facilitate left ventricular growth and improve function. We have recently completed a retrospective analysis of 70 attempts at fetal aortic valve dilation, of which 52 (70%) were technically successful.

We reached several conclusions:

1. Technically, successful aortic valve dilation has been possible in nearly ¾ of mid-gestation fetuses with aortic stenosis and factors predictive of progression to HLHS.
2. Shorter LV length is associated with technical failure, as is preprocedural imaging equivocal for aortic atresia.
3. There is a real risk of fetal hemodynamic instability (40%) and/or demise (13%).
4. Postprocedural aortic regurgitation is common (40%), is associated with higher balloon-to-annulus ratio, and improves postprocedure for unknown reasons.
5. The LV does not seem to grow significantly after successful aortic valve dilation but does support growth of other left heart structures, and this contribution appears to be unique to fetuses undergoing dilation.
6. Anatomic and physiologic features prior to intervention identify fetuses with the potential for biventricular outcome at birth. We developed a threshold score incorporating these features.

Based on this recent review, fetuses with LV long axis dimension Z-score of less than -2, fetuses with likely aortic atresia, and fetuses with threshold scores of less than 4 (on a scale of 5) are no longer considered to be routine candidates for fetal aortic valve dilation, based on their extremely low likelihood of achieving neonatal biventricular repair.

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CCT

Abstract Title: 3-D Echocardiography Current Status and Uses
Presenter: Gerald R. Marx, MD; Boston Children's Hospital, Boston, MA USA

Objective

1. Review clinical applications of transthoracic three-dimensional echo-cardiography for complex malformations including muscular ventricular septal defects, Ebstein's anomaly of the tricuspid valve, and conotruncal malformations such as double outlet right ventricle.
2. Review current status and clinical applications of three-dimensional trans-esophageal echocardiography.
3. Review current status and clinical applications of intra-operative epicardial three-dimensional echocardiography.
4. Review current status and clinical applications of three-dimensional echo-cardiography to measure ventricular volumes and mass and ejection fraction in congenital heart disease.

Three-dimensional echocardiography is used in a variety of clinical conditions, on a routine basis, at many echocardiographic laboratories. Such utilization provides imaging planes and projections in a three-dimensional format to add additional information to the two-dimensional echocardiography studies. The routine use is based on matrix array technology allowing for expedient three-dimensional echocardiographic imaging with relatively small pediatric transducers which are ideal for transthoracic imaging. Transthoracic three-dimensional echocardiographic imaging is ideal for understanding the spatial anatomy of complex atrial and ventricular septal defects and

complex conoltruncal malformations, such as double outlet right ventricle. Three-dimensional trans-esophageal echocardiography is routinely utilized in older children, adolescents and young adult patients. This imaging has been primarily employed in the operating room for mitral, aortic or tricuspid repairs, or for complex left ventricular outflow tract obstruction. In smaller patients, in whom the transesophageal echocardiographic probe is too large, epicardial three-dimensional echocardiographic imaging has been routinely applied to gain additional anatomic information. In addition to providing an enhanced understanding of complex congenital and valvar abnormalities, three-dimensional echocardiography can now allow for measurement of mass and volumes of the left, right and single ventricles in the infants, children and adolescents and adults with congenital heart disease.

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CCT

Abstract Title: Surgical Strategies to Maximize Neurological Outcomes

Presenter: Peter Pastuszko, MD; Division of Cardiovascular Surgery; Children's Specialist of San Diego; HS Associate Clinical Professor of Surgery, University of California, San Diego; San Diego, CA USA

Objective

To provide a better understanding of the techniques involved in congenital heart surgery aimed at reducing neurologic morbidity and give an overview of the evidence supporting their use.

Abstract

Many of the complex congenital heart defects were uniformly fatal 25 to 30 years ago. Today, the children undergoing surgical correction of these defects are looking at survival rates in the range of 80-90%. The tremendous improvements made in the care of these individuals cannot be understated. However, it is now well documented that many of these adolescents and young adults have significant neurologic morbidity, such as difficulty with visuo-spatial orientation, motor skills, as well as behavioral and academic problems. It has also been shown that surgery can only in part explain these neurological outcomes. Other elements such as genetic factors, fetal, pre- and post-operative hypoxia/ischemia and others are now known to play a significant role in the complex etiology of these neurological sequelae. Nonetheless, surgery, and in particular the use of cardiopulmonary bypass, have been and continue to be a focus of extensive research and debate. Over the past two decades, an understanding and general agreement has been reached as to what may be the appropriate levels of hemodilution, degree of hypothermia and pH management. Meanwhile, the use of deep hypothermic circulatory arrest (DHCA) continues to be challenged and alternative perfusion techniques, such as selective cerebral perfusion (SCP), have been advocated. Merits and limitations of these techniques, as well the potential direction of neuroprotection in the field of congenital heart surgery will be discussed.

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CCT

Read Part IV: "Abstracts from 'Evolving Concepts in the Management of Complex Congenital Heart Disease II' - in the July issue

Medical News, Products and Information

World First Remote Heart Operation to be Carried Out in Leicester Using Robotic Arm

A pioneering world first robotics system operation is to be conducted at Glenfield Hospital Leicester thanks to expertise at the University of Leicester and University Hospitals of Leicester.

Dr. André Ng, Senior Lecturer in Cardiovascular Sciences at the University of Leicester and Consultant Cardiologist and Electrophysiologist, Glenfield Hospital, University Hospitals of Leicester, is the first person in the world to carry out the operation remotely on patients using this system.

He will use the Catheter Robotics Remote Catheter Manipulation System for the first time in a heart rhythm treatment procedure.

The system is novel because it allows a doctor to carry out a common heart treatment procedure remotely using a robotic arm.

These procedures involve inserting thin wires, called catheters, into blood vessels at the top of the groin and advanced into the heart chambers. Electrodes on the catheters record and stimulate different regions of the heart to help the doctor identify the cause of the heart rhythm problem which usually involves an abnormality in the electrical wiring system of the heart. Once this area is identified, one of the catheters will be placed at the location to ablate or "burn" the tissue to cure the problem. Catheter ablation has been developed and used over the past two decades effectively in many patients suffering palpitations due to heart rhythm disturbances.

Dr. Ng said, "The new Robotic procedure is an important step forward because, while some procedures are straightforward, others can take several hours. Because X-rays are used to allow the doctor to monitor what is going on inside the patient, it means that doctors standing close to the patient wear radiation shields such as lead aprons which are burdensome. Protracted procedures can lead to clinician fatigue and high cumulative radiation exposure."

"The benefit of the Robotics system to the patient is that movement of the catheter could be done with great precision. It is anticipated that further developments of the system may allow complex procedures to be made more streamlined. On the other hand, benefits to the doctor are that heavy lead aprons would not be necessary as he / she will be controlling the movements of the catheter using the Remote Controller at a distance from the patient outside the radiation area and that he / she can be sitting closer to the monitors displaying electrical signals and x-ray images as opposed to standing at some distance across the room from them which is current practice."

Dr. Ng and his team's international standing and leading position in the management of heart rhythm disorders are reflected in the invitation to be the first to apply this new Robotics System in clinical procedures which also affirms the world-class research and pioneering work at the University of Leicester.

The Remote Catheter Manipulation System (RCMS, Catheter Robotics Inc., New Jersey) is a new system and Dr. André Ng, who has extensive experience in EPS procedures, has been selected to apply the system in human studies for the first time in the world. Two other remote navigation systems are commercially available but one uses a huge magnetic field to control a magnetic tip catheter whilst the other uses a large deflectable sheath to move the catheter. The RCMS has the benefit of using standard EPS catheters which can be dismounted and remounted onto the system with ease. The technology has obtained CE mark through rigorous bench safety testing and pre-clinical studies and has now arrived at a stage where it can be applied to clinical procedures.

Dr. Ng is an expert in the management of heart rhythm disturbances especially in catheter ablation and the use of mapping systems in such procedures. The Department of Cardiology at Glenfield Hospital is one of the largest Electrophysiology Centres in the UK performing over 600 EPS procedures every year. Dr. Ng has a distinguished research profile in investigations into cardiac electrophysiology and arrhythmia mechanisms, leading both non-clinical and clinical teams of talented researchers. At the cutting edge of scientific research and development, the innovative work in his group has been acknowledged with many accolades including Young Investigator and Da Vinci Awards. He is also Director of pan-European training programmes on advanced three-dimensional mapping systems and arrhythmia ablation.

It is planned to extend the use of the robotic system in research trials to be supported by the new Leicester Cardiovascular Biomedical Research Unit.

People With Common Heart Defect Also More Likely to Have Brain Aneurysms

A new study shows that people with a common heart defect may also be more likely to have brain aneurysms. The study was published May 4, 2010, in the print issue of *Neurology*®, the medical journal of the American Academy of Neurology (AAN).

Up to 2% percent of the population is born with the heart defect called a bicuspid aortic valve (BAV). The aortic valve allows blood to flow from the heart to the aorta. It normally has three flaps that open and close to regulate blood flow. In people with a bicuspid aortic valve, the valve does not develop fully during gestation and there are two flaps instead of three.

Some people with BAV never have any problems, but many develop narrowing or leakage of the aortic valve, especially as adults.

Recent research has shown that the artery problems with BAV may also occur in the brain, and that BAV may be a connective tissue disorder. Brain aneurysms are a weakening in a brain artery that causes a bulge in the artery.

"Since brain aneurysms are a treatable problem that can lead to death and disability if they rupture, we wanted to find out how common they are in people with BAV," said study author Wouter Schievink, MD, Director of Microvascular Neurosurgery at Cedars-Sinai Medical Center in Los Angeles, Calif.

For the study, 61 people with BAV were screened for brain aneurysms, along with 291 people who did not have BAV, but were undergoing scans for a suspected stroke or brain tumor during the same time period.

Six of the 61 people with BAV had brain aneurysms, or 9.8%, compared to three of the 291 people who did not have BAV, or 1.1%. Studies have shown that 0.5-2% of the general adult population has brain aneurysms.

"While more research needs to be done to confirm these results, these findings show a significant increased risk of brain aneurysms in people with bicuspid aortic valves," Schievink said.

Schievink said the heart defect has been shown to cluster in families, and screening is generally recommended for close family members of people diagnosed with bicuspid aortic valves.

Cedars-Sinai heart transplantation tip sheet

Physician scientists at the Cedars-Sinai Heart Institute are presenting new findings on heart transplantation rejection factors such as race and

gender, the effects of pre-transplant smoking and whether homeless organ donors put recipients at higher risk for complications. The presentations took place April 21–24 in Chicago during the 30th Annual Scientific Meeting of the International Society of Heart and Lung Transplantation.

"The International Society for Heart and Lung Transplantation Scientific Sessions bring together the world's experts in these fields which also includes basic science and mechanical circulatory assist devices," said Jon Kobashigawa, MD, director of the Cedars-Sinai Heart Institute heart transplant program. "This important dialogue and exchange of valuable information will lead to improved survival and quality of life of our patients."

An associate director of the Cedars-Sinai Heart Institute, Kobashigawa is a past president of the International Society for Heart and Lung Transplantation. His research has resulted in groundbreaking medical protocols, such as customized anti-rejection medications for transplant patients.

Cedars-Sinai Heart Institute researchers will make more than 30 presentations during the meeting, including:

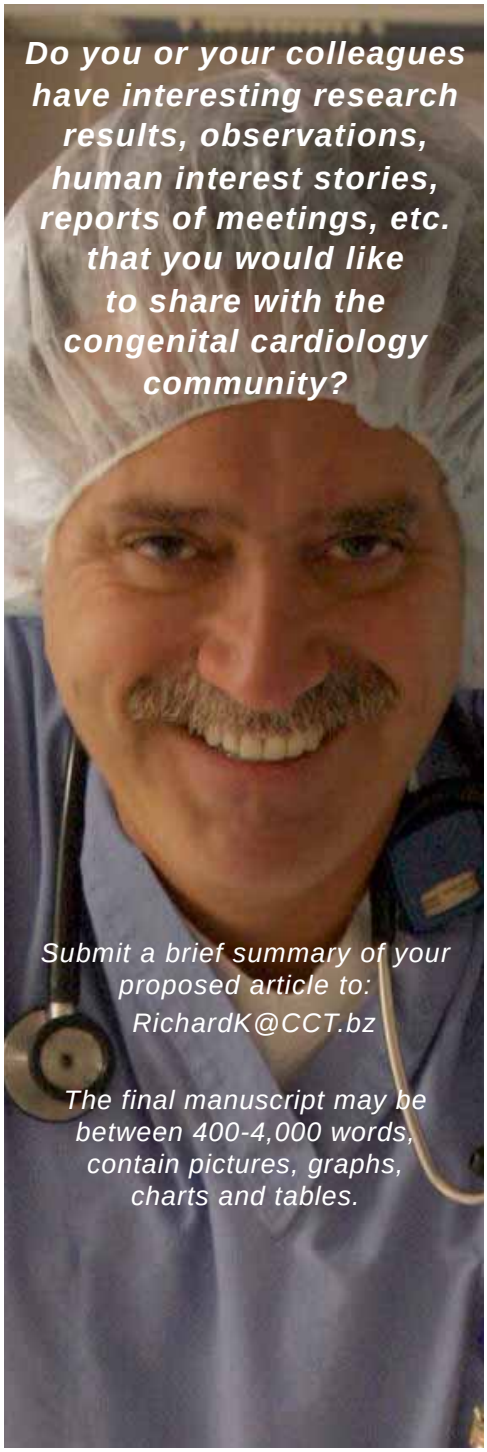
A review of 520 first-time heart transplant patients showed no significant difference survival rates based on age. Although patients 70 or older were more likely to have coronary artery disease and prior coronary bypass surgery, 10-year survival rates were equal to younger transplant patients. Older patients did have lower rates of diabetes mellitus, hypertension, obesity and smoking compared to the younger patients.

Transplant patients who receive hearts from organ donors who were homeless are at increased risk for complications and poor outcomes. Although homelessness is not included in the Centers for Disease Control criteria for high-risk organ donation, half the patients who received hearts from homeless organ donors died within the first year of transplantation. Researchers believe that homelessness leads to poor hygiene and increased susceptibility to infections, which can affect organ recipients. Further study is needed to confirm whether homelessness should be added to the high-risk organ donation category.

Patients who smoked prior to receiving a heart transplant experienced worse outcomes than patients who never smoked. Following transplant, 10% of patients who formerly smoked experienced non-fatal, major adverse cardiac events, such as a stroke or heart failure, compared to just 4 percent of transplant patients who never smoked.

After reviewing 10-year survival and outcomes of 574 transplant patients, researchers found no difference in outcome for patients who received a heart from a donor of a different race. Embargo lifts noon CDT, April 23.

Transplant patients who receive donor hearts from the opposite gender typically have worse outcomes than patients who receive a heart from the same gender. Males who received hearts from males had a 69% survival rate 10 years after transplant, compared to 59% 10 year survival rate for males who received hearts from females. Females who received hearts from females had a 71% survival rate, compared to 58% for females who received hearts from males.



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