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Medtronic Melody® Transcatheter Pulmonary Valve .... Improving the Quality of Life

By Sharon L. Hill, ACNP-BC, Joanne L. Chisolm, BSN, and John P. Cheatham, MD

Case Report

Clinical History

The patient is a 34 year old female with Down's Syndrome and complete AV septal defect, pulmonary atresia, and VSD who has been treated her entire life at C.S. Mott Children's Hospital, University of Michigan, at Ann Arbor. She had palliative bilateral modified Blalock-Taussig shunts (MBTS) as an infant and in 1987 had complete AV septal defect repair with RV-PA conduit, patch VSD closure, along with closure of both MBTS. Ten years later, the degenerating RV-PA conduit was replaced with a 24 mm Hancock porcine valve conduit. She had progressive conduit dysfunction with combined stenosis and moderate pulmonary regurgitation along with associated right heart failure with increasing tricuspid regurgitation and tricuspid stenosis. Her family stated that she could no longer walk up a flight of stairs at home, fell asleep spontaneously throughout the day, and had generalized edema on diuretic therapy. She was New York Heart Association Class III-IV. Her pre-catheterization MRI study confirmed conduit degeneration with combined stenosis and regurgitation with right pulmonary artery stenosis, decreased blood flow of 32% to the right lung and 68% to the left lung. She was referred for percutaneous pulmonary valve replacement using the Medtronic Melody® Transcatheter Pulmonary Valve (TPV).

Procedure

Under general endotracheal anesthesia and full heparinization, a right and left hemodynamic cardiac catheterization was performed with angiography. Right ventricular pressure was elevated at 75/0,19 with simultaneous left ventricular pressure of 90/0,12. Her PA pressure was 32/14 with mean of 24 in the left and 26/15 with mean of 20 in right. A right pulmonary artery (RPA) angiogram demonstrated significant RPA branch stenosis at the insertion site of the previous MBTS, involving the right upper and lower lobe

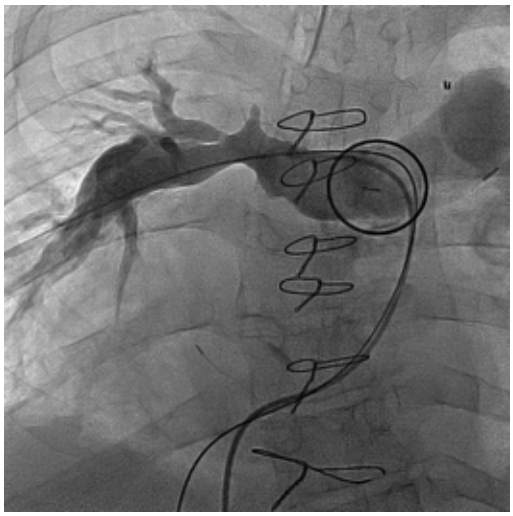

Figure 1. RPA angiogram shows RPA branch stenosis at the insertion site of the previous modified BT shunt.

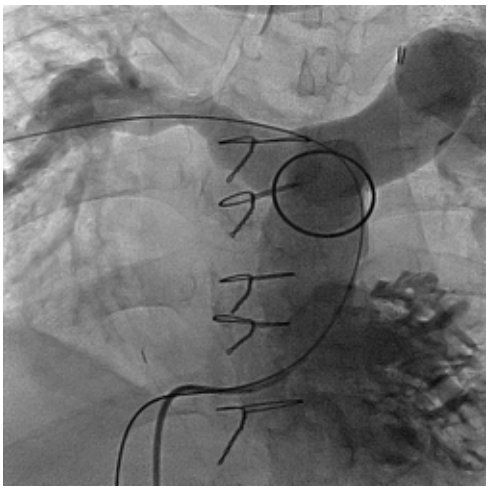

Figure 2. Pulmonary artery angiogram shows moderate pulmonary regurgitation with calcification of the conduit.

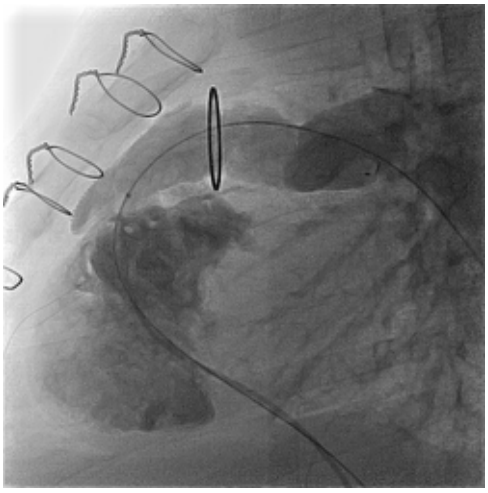

Figure 3. RVOT angiogram shows a dilated and hypertrophied right ventricle with calcified and stenotic Hancock conduit.

branches (Figure 1). This area measured 7-8 mm in diameter, whereas, the rest of the pulmonary artery measured 16 mm. There was moderate pulmonary regurgitation with calcification of the conduit (Figure 2). An angiogram performed in the right ventricular outflow tract (RVOT) showed a dilated and hypertrophic right ventricle with an unusual attachment of the calcified Hancock conduit with almost a hood in the RVOT and some narrowing (Figure 3). The narrowest diameter of the conduit measured 16.3 mm. The aortic root angiogram demonstrated normal coronary arteries which appeared remote

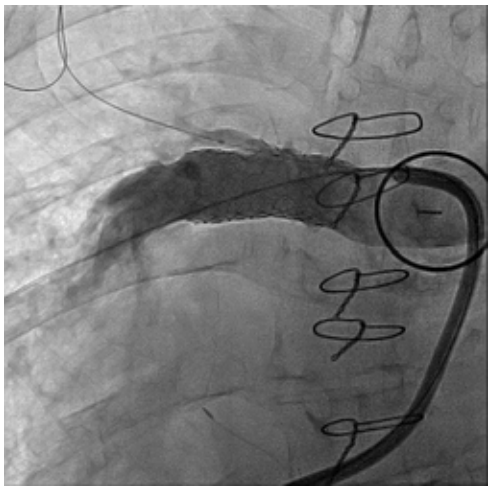

Figure 4. Repeat RPA angiogram s/p implantation with a 36mm long ev3 Max LD stent and balloon angioplasty of the stenotic right upper lobe branch using a 6mm coronary balloon catheter through the open cells of the RPA stent.

from the titanium ring of the valve conduit, therefore little risk for coronary artery compression related to the planned procedure. There was mild aortic insufficiency.

The RPA branch stenosis was treated with a 36 mm long ev3 Max LD stent hand-crimped on a 16 mm X 4 cm BIB delivery catheter followed by redilation using an ATLAS balloon catheter with complete expansion. A repeat angiogram in the RPA demonstrated relief of obstruction. A 6 mm coronary balloon was used to dilate the proximal stenosis of the right upper lobe branch through the open cells of the RPA stent (Figure 4).

Compliance testing of the calcified conduit was performed followed by implantation of the Melody® TPV using the Ensemble Balloon Delivery System as per protocol (Figure 5). A Kevlar-coated ATLAS® balloon catheter was used to completely expand the the stent to 20 mm (Figure 6). Pulmonary artery (PA) pressure recording post implant revealed no significant gradient across the Melody® Valve. There was a 10 mmHg peak systolic gradient across the entry site or attachment of the hooded insertion of the RVOT. Ending RV pressure was 40/4,17 with an LV pressure of 85/4,15. A follow-up PA angiogram using the Multi-Track® angiographic catheter demonstrated no pulmonary regurgitation with excellent positioning of the valve and complete relief

"The patient is a 34 year old female with Down's Syndrome and complete AV septal defect, pulmonary atresia, and VSD.... She had palliative bilateral modified Blalock-Taussig shunts (MBTS) as an infant and in 1987 had complete AV septal defect repair with RV-PA conduit, patch VSD closure, along with closure of both MBTS. Ten years later, the degenerating RV-PA conduit was replaced with a 24 mm Hancock porcine valve conduit. She had progressive conduit dysfunction with combined stenosis and moderate pulmonary regurgitation along with associated right heart failure with increasing tricuspid regurgitation and tricuspid stenosis."

of the RPA stenosis with brisk flow into the right and left pulmonary artery branches (Figure 7). An RVOT angiogram was also performed which demonstrated no significant residual stenosis except at the insertion site of the conduit within the RV muscle where there was a 10 mm gradient (Figure 8). An intracardiac echo advanced to the RV outflow tract showed coaptation of the Melody® Valve leaflets and no significant pulmonary regurgitation and a mean gradient of 4-5mmHg (Figure 9). Finally, a "down the barrel" angulation of the AP camera and straight lateral view was recorded as baseline comparison for future images (Figure 10).

At six month follow-up, the patient has improved to New York Heart Association

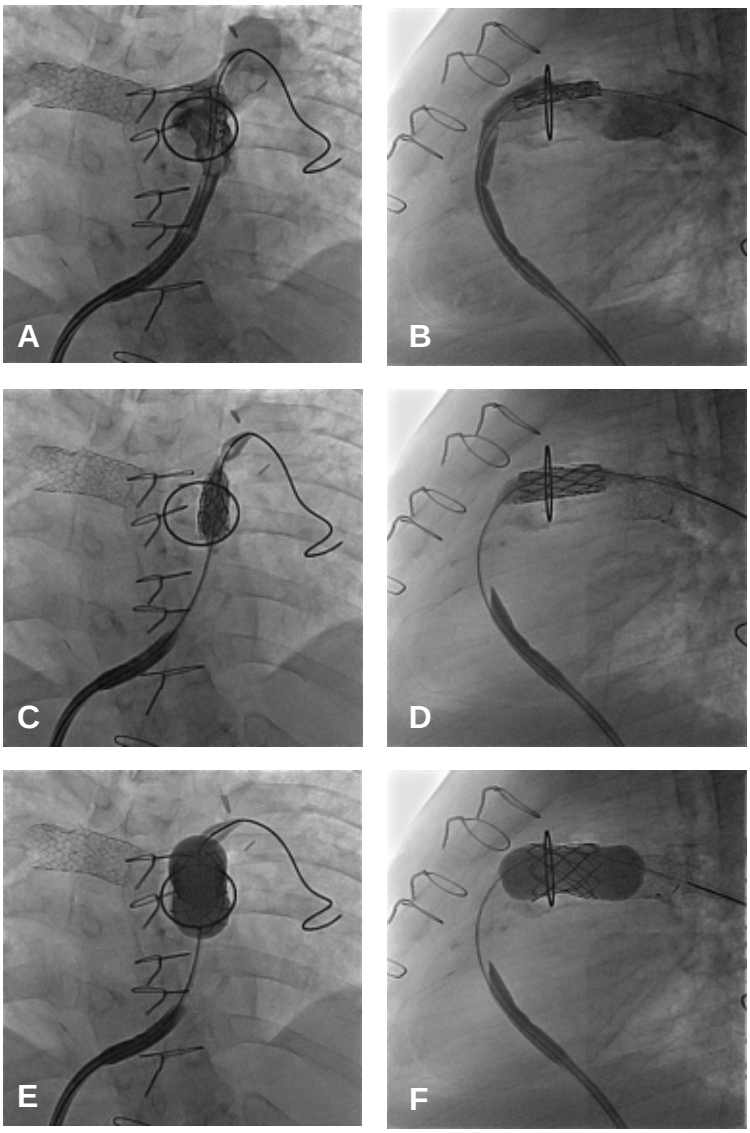

Figure 5. Hand injection for placement of the Melody TPV and Ensemble Balloon Delivery System across the titanium ring in Hancock conduit in AP & Lateral projection (A&B). AP and lateral projections of the inner balloon (C&D) then outer balloon (E&F) of the BIB catheter expanded to 4 atmospheres of pressure.

Class I. Imaging studies demonstrate no change in valve function and no evidence of stent fracture, with 13 mean gradient and no pulmonary regurgitation. The family reports the patient's activity level is markedly increased without any evidence of lethargy. She is asymptomatic and now riding her bicycle, climbing multiple flights of stairs, and walking long distances. She has regained the ability to dance, which she loves and has missed. Her attending cardiologist in Ann Arbor, who has followed her for over 30 years, calls the procedure "a miracle!"

Discussion

Bonhoeffer, et al (2000) reported the first percutaneous transcatheter bovine jugular valve implantation in a conduit in a 12 year old boy with stenosis and insufficiency of a prosthetic RV-PA conduit. Since then, patients with degenerating RV-PA conduits and RV dysfunction have successfully undergone percutaneous transcatheter pulmonary valve implantation using the Medtronic Melody® TPV under CE Mark in Europe and Canada, and more recently under a Food and Drug Administration (FDA) sponsored protocol in the United States (Lurz et al, 2009; Zahn et al, 2010). Recently, a Humanitarian Device

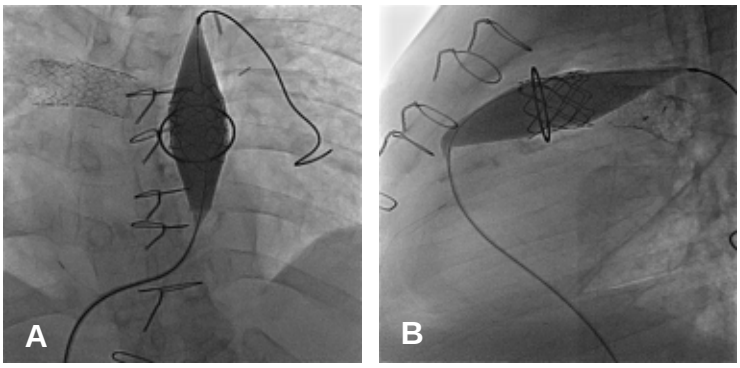

Figure 6. A 20mm x 2cm long Kevlar coated ATLAS balloon catheter inflated to 18 atmospheres of pressure with complete expansion in AP (A) and Lateral (B) view.

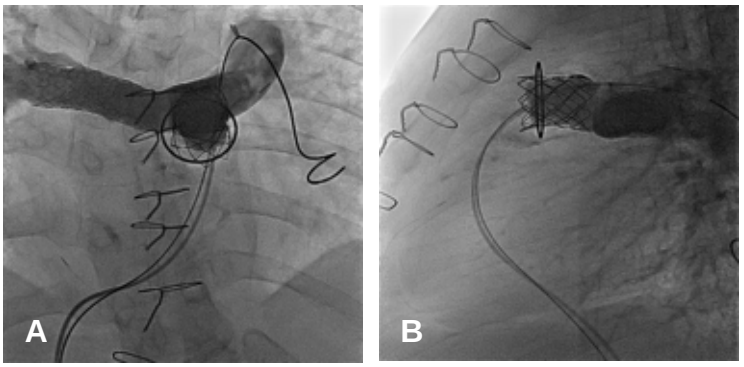

Figure 7. Post PA angiogram in the AP (A) and Lateral (B) projection using a Multi-Track angiographic catheter demonstrated no pulmonary artery regurgitation, relief of the RPA stenosis with brisk flow into the right and left pulmonary artery branches.

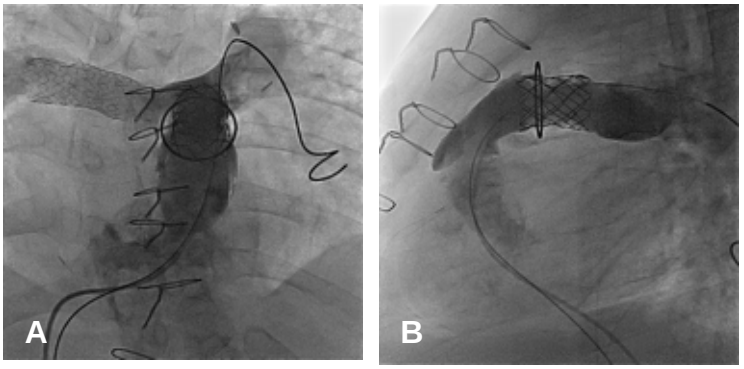

Figure 8. Post RVOT angiogram in the AP (A) and Lateral (B) projection using a Multi-Track angiographic catheter shows the area of mild stenosis (10mmHg gradient) at the insertion site of the conduit within the RV muscle, otherwise no significant residual stenosis.

Exemption (HDE) was granted. This is a special type of regulatory approval by the FDA for medical devices which have proven safety and probable benefit, and intended for fewer than 4,000 patients per year. More than 1200 patients in 90 centers worldwide have undergone transcatheter pulmonary valve replacement using the Medtronic Melody® TPV (personal communication, Philipp Bonhoeffer MD, March 2010). This non-surgical option offers an alternative treatment for congenital heart disease patients who have a degenerative RV-PA conduit. Transcatheter pulmonary valve replacement may decrease the number of open heart surgeries needed over the course of a lifetime for many patients. This new interventional procedure can offer patients, who were previously considered inoperable, a competent pulmonary valve with relief of

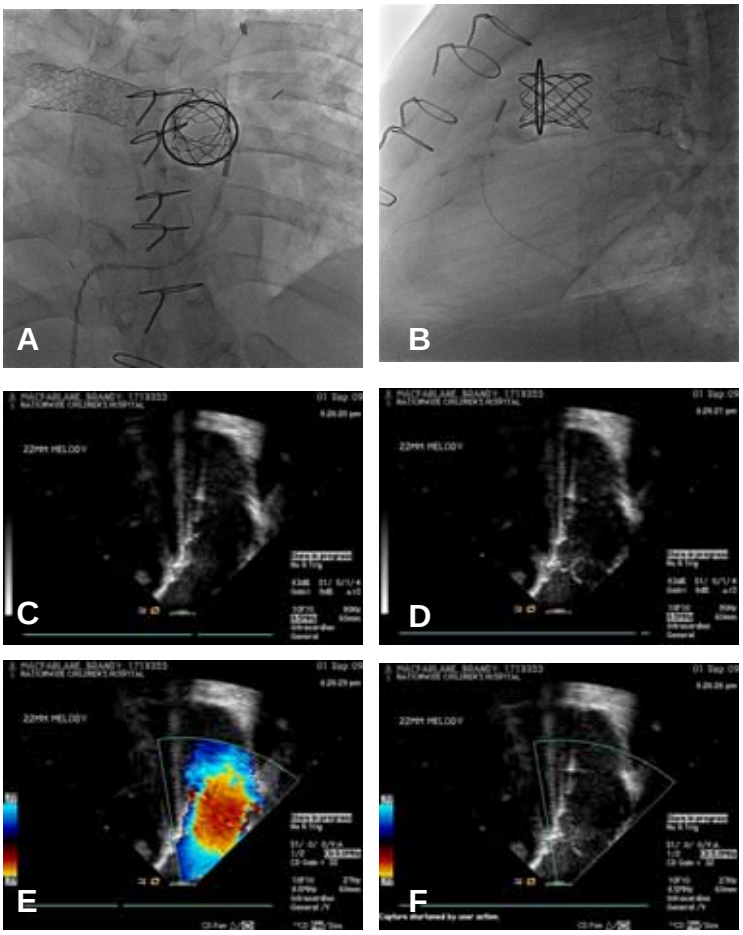

Figure 9. (A&B) Shows placement of the ICE catheter in the RVOT beneath the Melody TPV. ICE imaging shows the valve in the open (C) and closed (D) position with coaptation of the valve leaflets. Color flow Doppler in the open (E) and closed (F) position shows no significant pulmonary regurgitation and a mean gradient of 4-5mmHg.

stenosis and regurgitation, as well as improved quality of life. The patients are happy and grateful to regain the ability to perform the activities they love (Figure. 11).

Special Acknowledgement: Macdonald "Mac" Dick II, MD and Aimee K. Armstrong, MD; C.S. Mott Children's Hospital.

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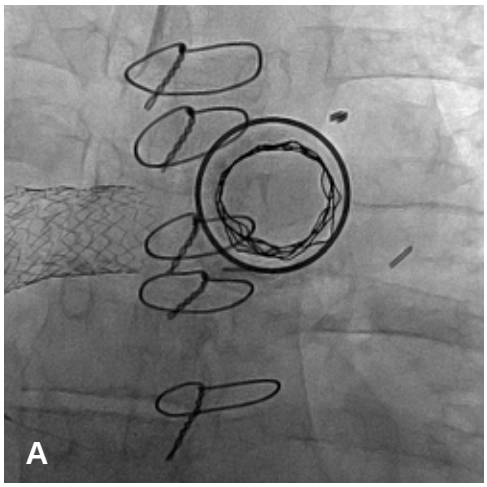

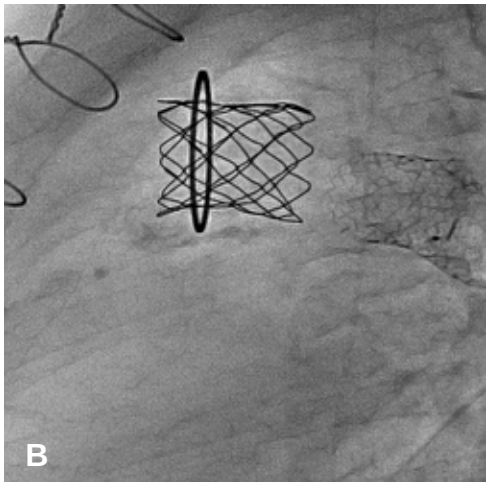

Figure 10. (A) "Down the barrel" and (B) Lateral view post procedure.

Figure 11. Happy patient… returned to dancing and all other activities!

"This new interventional procedure can offer patients, who were previously considered inoperable, a competent pulmonary valve with relief of stenosis and regurgitation, as well as improved quality of life."

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Highlights from "The Evolving Concepts in the Management of Complex Congenital Heart Disease II:"

January 14-16th 2010; San Diego, California; Sponsored by Rady Children's Hospital—San Diego and UCSD; Coordinators John Lamberti and John Moore

By John W. Moore, MD, MPH

Our second symposium entitled *Evolving Concepts in the Management of Complex Congenital Heart Disease,* took place in mid-January at the Hyatt Regency Mission Bay Spa and Marina in San Diego.

The first symposium occurred in October 2007, and was a two-day program focusing on surgical management of difficult defects and peri-operative care. Our second symposium was held in mid-January 2010, allowing many attendees to escape harsh winter conditions, stay at a nice bay-side resort and enjoy San Diego's mild Mediterranean climate and many family-oriented attractions. Furthermore, the program was expanded to three days and many sessions were added to provide the audience with a comprehensive cardiology and cardiac surgery program. There were approximately 200 attendees coming from most of the United States; the majority were practicing pediatric cardiologists.

Program Sessions included: Office Cardiology; Cardiomyopathy and Pulmonary Hypertension; Imaging; Electrophysiology; Catheterization; Hypoplastic Left Heart Syndrome; Difficult Surgical Problems; Valvular Heart Disease; and Adult Congenital Heart Disease. The keynote address was given by Jeff Jacobs on the morning of the final day. He discussed the Status and Utility of Clinical Data Bases from the perspectives of quality improvement, research and health care administration.

The Office Cardiology Session featured Victoria Vetter, Lloyd Tani, and Jane Burns. Victoria Vetter discussed ADHD in patients with and without congenital heart disease. She discussed the role of ECG's and the use of stimulant medication. Lloyd Tani provided a case-based discussion of the management of children with aortic and mitral regurgitation. He gave thoughtful advice about the role of medical therapy and the timing of surgery. Jane Burns informed the audience about the current status of Kawasaki Disease research and the latest recommendations for therapy.

Rose Pavillion at Rady Chidren's Hospital - San Diego, CA

Table at the Faculty Dinner with faculty members: Beth Printz, Joel Kirsch, Howaida El Said, and Jane Burns

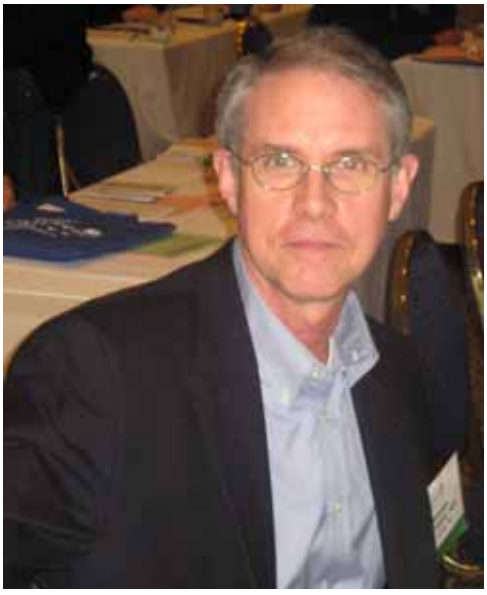
The next session dealt with Cardiomyopathy and Pulmonary Hypertension. Speakers included Steven Lipshultz, Daniel Bernstein, and Dunbar Ivy. Steven Lipshultz discussed the Pediatric Cardiomyopathy Registry, and made recommendations for the evaluation and medical treatment for cardiomyopathy patients. A complete discussion of devices for treatment of pediatric heart failure patients and of pediatric cardiac transplantation including the results, patient management and complications encountered after OHT was given by Daniel Bernstein. Dunbar Ivy provided a thorough review of the evaluation and the pharmacological management of pulmonary hypertension.

The Imaging Session was kicked off by Gerald Marx who showed some fantastic 3-D echocardiograms and discussed their clinical utility. Beth Printz showed some cardiac MRI's and CT's and described her considerations in choosing one or the other modality for imaging. Lloyd Tani returned to the podium to discuss the role of routine 2-D and Doppler echocardiography in the evaluation of mitral and aortic regurgitation.

George Van Hare led the Electrophysiology Session. Joel Kirsh discussed Sudden Death and the Channelopathies. George Van Hare laid out treatment for native and post-operative Atrial Ectopic Tachycardia, and Junctional Ectopic Tachycardia. Kevin Shannon

Jenny Boyd and Victoria Vetter

Gary Webb and John Child



George Van Hare

concluded the session with a practical discussion of Device Therapy for Tachyarrhythmia and Heart Failure. He talked about the indications for Bi-Ventricular Pacing and for AICD in children and in adults with congenital heart disease.

James Perry

John Lamberti and Joseph Dearani

John Moore and Anjan Batra

The Catheterization Session featured Zahid Amin, Audrey Marshall, and Howaida El Said. Zahid Amin spoke about device closure of VSD, both percutaneous and per-ventricular. He hinted about some new device designs which are still in the drafting stages. Audrey Marshall provided an excellent review of Fetal Interventional Cardiology, including discussions of aortic and pulmonary valvuloplasty, and PFO enlargement and

James Lock and Scott Kiewer

stenting. Howaida El Said discussed the problems involved in organizing and running a Hybrid program for Hypoplastic Left Heart Syndrome. In Dr. El Said's program hybrid palliation is only offered to high-risk patients. Her talk provided an excellent segue to the Hypoplastic Left Heart Syndrome session which followed immediately thereafter.

The Hypoplastic Left Heart Syndrome (HLHS) Session featured speakers John Cheatham who described the Columbus experience in which Hybrid palliation is offered to all patients with HLHS, and James Tweddell who agrued "It's Hard to 'Top' a Well-Done Norwood." Thomas Spray followed with a comprehensive review of what happens after stage I and II procedures in patient with HLHS. He concluded that Fontan completion in this patient group is similar to the single ventricle group at large. A lively panel discussion followed this Session.

Following HLHS, the next Session dealt with Managing Difficult Surgical Problems. Frank Hanley discussed his ground-breaking approach to Management of Patients with One or Two Ventricles and MAPCAS rather than true pulmonary arteries. He also gave Mohan Reddy's talk about the surgical management of the Premature Infant with Complex Congenital Heart Disease. Next, Peter Pastuszko discussed Surgical Strategies to maximize neurological Outcomes after Cardiac Surgery, and Gil Wernovsky gave a very thoughtful talk about the impact of Patient-related, Procedure-related and ICU-related Factors on Long-Term Quality of Life.

On the last day, the first Session dealt with important topics in Valvular Heart Disease. Vaughn Starnes discussed his experience with the Infant Ross/Konno Procedure. James Lock outlined a comprehensive strategy for Incorporating the Percutaneous Pulmonary

"Program Sessions included: Office Cardiology; Cardiomyopathy and Pulmonary Hypertension; Imaging; Electrophysiology; Catheterization; Hypoplastic Left Heart Syndrome; Difficult Surgical Problems; Valvular Heart Disease; and Adult Congenital Heart Disease."

Valve into a Long-Term Patient Care Strategy. Joseph Dearani gave a very clear and comprehensive presentation about Repair of Ebstein's Anomaly of the Tricuspid Valve.

The Final Session covered Adult Congenital Heart Disease. Gary Webb described the challenges involved in establishing an ACHD Program. John Child provided a sweeping case-based discussion of patient care challenges. And, with very little audience attrition, Anjan Batra gave the last talk in which he summarized the management of arrhythmias in this patient population.

Evolving Concepts II adjourned with the satisfied applause of the attendees, and with very positive reviews. We plan to reconvene in January of 2012 with a similar program. Join us for a pleasant and fruitful break from winter at *Evolving Concepts in the Management of Complex Congenital Heart Disease III!*

CCT

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Congenital Cardiology Today will be publishing many of the abstracts from *Evolving Concepts in the Management of Complex Congenital Heart Disease II* in this and the next few issues.

The topics and presenters are:

- How Far Can We Go With VSD Closure? *by Zahid Amin, MD*
- Evolving Concepts in the Management of Arrhythmias in Adults with Congenital Heart Disease *by Anjan S Batra, MD*
- Pediatric Cardiac Transplantation: The Next 25 Years *by Daniel Bernstein, MD*
- Kawasaki Disease Update *by Jane C. Burns MD*
- Assessment of the Hybrid Approach to the Management of HLHS *by John P. Cheatham, MD*
- Late Challenges in Adult CHD – A Case-Based Approach *by John S. Child, MD*
- Surgery for Ebstein's Anomaly *by Joseph A. Dearani, MD*
- Starting a Hybrid Program *by Howaida G. El-Said, MD*
- Management Of Patients With Aorto-Pulmonary Collaterals *by Frank L Hanley,* MD
- Update on Medical Management of Severe Pediatric Pulmonary Hypertension *by D. Dunbar Ivy, MD*
- 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
- When MRI, When CT *by Beth Feller Printz, MD, PhD*
- Indications for AICD, Anti-Tachycardia PM, Bi-V Pacing *by Kevin Shannon, MD*
- Is the Fontan Procedure Different in HLHS versus Other Single Ventricles? *by Thomas L. Spray, MD*
- The Infant Ross / Konno: Early and Late Results *by Vaughn A. Starnes, MD*
- Management of Children with Aortic or Mitral Regurgitation: Role of Medical Therapy and Timing of Surgery *by Lloyd Y. Tani, MD*
- Role of Echocardiography in the Evaluation of Mitral and Aortic Regurgitation *by Lloyd Y. Tani, MD*
- It's Hard to Top a Well-Done Norwood *by James S Tweddell, MD*
- Treatment of AET and JET – Native and Post-Op *by George F. Van Hare, MD*
- DHD in Patients with and without CHD, Use of Stimulant Medication *by Victoria L. Vetter, MD, MPH*
- Establishing an ACHD Program *by Gary Webb, MD*

JUNE 2010
MEDICAL MEETING FOCUS

20th Annual Congenital Heart Disease in the Adult
June 6-9, 2010; Stevenson, WA USA
www.ohsu.edu/xd/education/schools/school-of-medicine/gme-cme/cme/ACHD.cfm

Sponsored by the Oregon Health & Science University

Program Co-Directors: David J. Sahn, MD, MACC, Oregon Health & Science University, Portland, OR; Erwin Oechslin, MD, FRCP(C), FESC, University of Toronto, University Health Network, Toronto, ON, Canada; Gary D. Webb, MD, FRCP(C), FACC, Cincinnati Adolescent and Adult Congenital Heart Center, Cincinnati, OH; Craig S. Broberg, MD, FACC Oregon Health & Science University Portland, OR; Pamela D. Miner, RN, MN, NP, UCLA Adult Congenital Heart Disease Center Los Angeles, CA

Meeting will present a comprehensive review of problems and issues related to the more common forms of congenital heart disease (CHD) in the adult, including:

- Methods of Diagnosis
- Methods of Treatment
- Career and Lifestyle Counseling
- Pregnancy – Care and Counseling
- Latest Large Multicenter Studies Related to Natural History of CHD in Adults and Treatment Outcomes
- Postoperative Residua of Congenital Disease

Sessions are oriented to stress the management and physiology of unoperated and post-operative CHD. Small group breakout sessions with the faculty to foucs on problem cases brought by the faculty or by the participants.

Selected List of Faculty: Jamil Aboulhosn, MD; John Deanfield, MB, FACC; Michael A. Gatzoulis, MD, PhD, FACC; Welton Gersony, MD; Michelle Gurvitz, MD, FACC; Timothy Hornung, MBBChir, MRCP; Michael Jerosch-Herold, PhD; Michael J. Landzberg, MD, FACC; Alison Meadows, MD; Barbara J.M. Mulder, MD; Joseph K. Perloff, MD, FACC; Carsten Rickers, MD; Samuel C.B. Siu, MD, SM, FRCP (C), FACC; Thomas Spray, MD, FACC; Karen Stout, MD, FACC; Judith Therrien, MD, FRC (C); Gruschen Veldtman, MBChB MRCP (UK); Gil Wernovsky, MD, FACC; Jack M. Colman, MD, FRCPC, FACC; Andrew Crean, MD, MRCP, FRCR; Louise Harris, MD, ChB, FACC; Eric M. Horlick, MD, FRCPC; Adrienne Kovacs, PhD; Andrew N. Redington, MD, FRCPC; Candice Silversides, MD, MS, FRCP(C); Glen Van Arsdell, MD, FRCSC; Laurie Armsby, MD; Seshadri Balaji, MD, MRCP, FACC; Dianna M. Bardo, MD; Grant H. Burch, MD, FACC; Sanjiv Kaul, MD, FACC; Stephen M. Langley, MD, FACC; Brian J. Morrison, MD, FACC; Joseph Weiss, MD, PhD; and others.

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Highlights from "The 18th Utah Conference on Congenital Cardiovascular Disease"

By Collin G. Cowley, MD

The *18th Utah Conference on Congenital Cardiovascular Disease* convened in the mountainous beauty of Snowbird, Utah February 21-24, 2010. Faculty from across the United States and Australia gathered to present their work and lead discussions on a variety of issues important to the care of children and adults with heart disease.

This year's keynote speaker was Dr Pedro del Nido of Boston Children's Hospital, who presented an excellent and provocative talk on recruitment of the borderline left ventricle. The conference was again organized over a four-day period allowing time during the middle of each day for participants to enjoy the scenic and recreational opportunities unique to this area. A summary of the speakers and their presentations is included (Table below).

"This year's keynote speaker was Dr. Pedro del Nido of Boston Children's Hospital who presented his work on recruitment of the borderline left ventricle."

18th Utah Conference on Congenital Cardiovascular Disease – Topics and Speakers	
Recruiting the borderline left ventricle	Dr. Pedro del Nido
Optimal methods of preoperative imaging	Dr. Shaji Menon
Surgical strategies for small neonates	Dr. Aditya Kaza
Ventilation strategies for CHD - options and outcomes	Dr. Ira Cheifetz
Perioperative monitoring of cerebral oxygenation	Dr. Madolin Witte
Neuroprotection of neonates with CHD	Dr. Patrick McQuillen
Perventricular VSD Closure	Dr. Geoff Lane
New Technology and the Pulmonary Conduit	Dr. John Hawkins
Advances in the Invasive Treatment of Pediatric Rhythm Disorders	Dr. John Triedman
Transcatheter Interventions For Structural Heart Disease In Adults	Dr. John Whisenant
Anticoagulation in Children - Are We Missing the Clot?	Dr. Gordon Mack
Fetal Diagnosis of CHD - Impact and Outcomes	Dr. Nelangi Pinto
PFO, Strokes and Migraine Headaches in the Pediatric Population	Dr. Rachel McCandless
What's New in Pediatric Heart Transplantation	Dr. Melanie Everitt
Neuropsychological Problems in Children with Heart Disease - Part I	Dr. Richard Martini
Neuropsychological Problems in Children with Heart Disease - Part II	Dr. Lisa Giles
Debate: Comfort Care for HLHS should be offered	Dr. Mike Puchalski
Debate: Comfort Care for HLHS should not be offered	Dr. Phil Burch
Functional Outcomes in Children After Single Ventricle Palliation	Dr. Rick Ohye
Surgical Management of the Failing Fontan Patient	Dr. Constantine Mavroudis
Genetics of Arrhythmias	Dr. Martin Tristani
Advances in our understanding of genotype and phenotype	Dr. Cammon Arrington
Update on Animal Models of CHD	Dr. Deborah Frank
The Science and Practice of Preventing Congenital Heart Defects	Dr. Lorenzo Botto

Dr. Cammon Arrington presents his cutting-edge research on advances in our understanding of genotype and phenotype.

University of Utah Pediatric Cardiology faculty Drs. Rachel McCandless and Nelangi Pinto enjoying themselves at the faculty dinner.

Dr. John Triedman celebrates his winning form after demonstrating his snowboard prowess.

Dr. Geoffrey Lane enjoys a cold one at the banquet while listening to some excellent jazz music.

Rick Ohye enjoys a moment with his very fast daughters, Maddie and Lily.

This year's meeting again included a variety of social events with great food and live music and the entertainment of the ski race that has increasingly become the forum for resolution of debates from earlier in the meeting.

The 19th Utah Conference on Congenital Cardiovascular Disease will be held in February 2012 with plans to feature international experts in the field.

The conference was presented by the Department of Pediatrics, University of Utah School of Medicine and primary CHildren's Pediatric Education Services.

Visit the website later in the year for further information: http://intermountainhealthcare.org/hospitals/primarychildrens/classes/classesformedical/conferences/Pages/home.aspx

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JULY 2010
MEDICAL MEETING FOCUS

PEDIATRIC & ADULT INTERVENTIONAL CARDIAC SYMPOSIUM
(PICS & AICS 2010)
With Live Case Demonstrations
July 18-21, 2010; Chicago, IL
www.picsymposium.com

Program DIrectors: Ziyad M. Hijazi, MD, MPH; William E. Hellenbrand, MD; Carlos Pedra, MD; John P. Cheatham, MD

Co-Directors: Zahid Amin, MD; Ted Feldman, MD

Guest Faculty: Teiji Akagi, MD; BG Alekyan, MD; Wail Alkaskhari, MD; Mazeni Alwi, MD; Emile Bacha, MD; David Balzer, MD; John Bass, MD; Lee Benson, MD; Farhouch Berdjis, MD; Felix Berger, MD; Jacek Bialkowski, MD; Philipp Bonhoeffer, MD; Robert Bonow, MD; Elchanan Bruckheimer, MD; Qi-Ling Cao, MD; Mario Carminati, MD; Alpay Celiker, MD; Jae Young Choi, MD; Roberto Cubeddu, MD; Bharat Dalvi, MD; Pedro Del Nido, MD; Carol Devellian, BS; Makram Ebeid, MD; Peter Ewert, MD; Horacio Faella, MD; Craig Fleishman, MD; Simone Fontes-Pedra, MD; Thomas Forbes, MD; Olaf Franzen, MD; Yun Ching Fu, MD; M. Omar Galal, MD; Mark Galantowicz, MD; Ricardo Gamboa, MD; Wei Gao, MD; J V De Giovanni, MD; Miguel Granja, MD; Donald Hagler, MD; Ali Halabi, MD; Sharon Hill, ACNP; Ralf Holzer, MD; Eric Horlick, MD; Reda Ibrahim, MD; Michel Ilbawi, MD; Frank Ing, MD; Ignacio Ingelessis, MD; Mansour Al Joufan, MD; Thomas Jones, MD; Saibal Kar, MD; Cliff Kavinsky, MD; Jan Kovac, MD; Seong-Ho Kim, MD, PhD; Terry D. King, MD; Charles S. Kleinman, MD; Krishna Kumar, MD; Geoffrey Lane, MD; Larry Latson, MD; Trong-Phi Le, MD; Daniel Levi, MD; Achiau Ludomirsky, MD; Raj Makkar, MD; Gerard Martin, MD; Jozef Masura, MD; Tarek Momenah, MD; John W. Moore, MD; Phillip Moore, MD; Michael Mullen, MD; Charles E. Mullins, MD; Toshio Nakanishi, MD; Kathleen Nolan, (RT) (R), (CV); David Nykanen, MD; Eustaquio Onorato, MD; Shakeel A. Qureshi, MD; David Reuter, MD; John Rhodes, MD; Richard Ringel, MD; Jonathan Rome, MD; Russsell D'Sa, CVT; Masood Sadiq, MD; Girish Shirali, MD; Terry Sideris, MD; Horst Sievert, MD; Dietmar Schranz, MD; Frank Silvestry, MD; Sherm Sorensen, MD; Jonathan Tobis, MD; Zoltan Turi, MD; Mike Tynan, MD; Julie Vincent, MD; Robert Vincent, MD; Kevin Walsh, MD; Jou-Kou Wang, MD; John Webb, MD; Howard Weber, MD; Paul Weinberg, MD; Gil Wernovsky, MD; Robert White, Jr, MD; James L. Wilkinson, MBBS; Neil Wilson, MD; Carlos Zabal, MD; and Evan Zahn, MD

Meeting Overview:

The meeting will feature live case demonstrations from multiple international centers demonstrating the latest in medical device technology using approved and investigational devices/valves/stents etc. This symposium will give the attendees an opportunity to interact with the faculty during lectures, interactive discussions, workshops, live case demonstrations and daily breakout sessions.

Special sessions will provide an in-depth focus on septal defect closure (PFO, ASD, VSD and PDA) using all available devices (approved and investigational), embolization therapies, coarctation stenting, aortic and mitral valve disease and their management in both children and adults, RVOT and pulmonic valve disease (featuring the new percutaneous valve implants with live cases of both the Melody and Edwards valves), hybrid intervention for Hypoplastic Left Heart Syndrome and muscular VSDs, stent/balloon angioplasty of branch pulmonary arteries, and "how to session," etc. There will be a special session on left atrial appendage, percutaneous aortic and mitral valve therapies presented by the experts in these fields.

There will be "Hot Debates" featuring a cardiologist and a cardiac surgeon debating forms of management and outcome, and the ever-popular session "My Nightmare Case in the Cath Lab."

This year there will be a special one-day seminar entitled, "Imaging in Congenital & Structural Cardiovascular Interventional Therapies," immediately preceding PICS ~ AICS. For information and program details visit the website.

Abstracts from "Evolving Concepts in the Management of Complex Congenital Heart Disease II" - Part 1

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

- How Far Can We Go With VSD Closure? *by Zahid Amin, MD*
- Evolving Concepts in the Management of Arrhythmias in Adults with Congenital Heart Disease *by Anjan S Batra, MD*
- Pediatric Cardiac Transplantation: The Next 25 Years *by Daniel Bernstein, MD*
- Kawasaki Disease Update *by Jane C. Burns MD*
- Assessment of the Hybrid Approach to the Management of HLHS *by John P. Cheatham, MD*

See Part II in the May issue of Congenital Cardiology Today

Abstract Title: How Far Can We Go With VSD Closure?
Presenter: Zahid Amin, MD; Professor of Pediatrics; Director, Cardiac Catheterization and Hybrid Suites, RUCH Center for Congenital and Structural Heart Disease; RUSH Medical Center, Chicago, IL US

Objective

The objectives of this presentation are to provide a brief history of VSD closure, followed by innovations in minimally invasive VSD closure from trans-catheter, robotic to perventricular procedures. The different types of device and delivery sheaths available and those that may become available will also be discussed.

Abstract

Ventricular septal defects (VSD) are the most common congenital cardiac lesions. VSD was the first defect that underwent closure with open heart surgery by Dr. Lillehei. From surgical perspective, it has become a relatively straightforward procedure. The most common type of VSD is perimembranous (or cono-ventricular) VSD and comprise nearly 90 % of all VSD. A small proportion of the patients have muscular VSD. Over the last several years, the interventional cardiologists have had the opportunity to close perimembranous and muscular VSD with the help of devices. Currently there are two devices approved for muscular VSD closure-CardioSEAL and Amplatzer MVSD device. There is no device approved for perimembranous VSD closure.

From a surgical standpoint, muscular VSD are difficult to close in the operating room as they may be embedded in the trabecular ventricular septum and the rims are not seen clearly. Patch closure, although successful, may not completely close the defect, may increase the cardiopulmonary bypass time and its associated morbidity and, increase chances of re-operation in cases of significant residual shunt.

Device closure of these defects has gained popularity as the procedure can be performed without cardiopulmonary bypass with decrease post-procedure morbidity.

At the current time, there are two options available to the interventionalist for VSD closure; transcatheter or perventricular closure. Both of these techniques are used on a routine basis and have

advantages over the surgical procedure-mainly no cardiopulmonary bypass (CPB).

In the data submitted to the FDA for muscular VSD device approval by AGA Medical Corporation, the risk of complications of transcatheter procedure increased significantly when the patient's weight dropped below 5.2 kg. Hence patients who are less than 5 kg are best-served by perventricular approach. This is a Hybrid approach in which the surgeon and the interventionalist are equally involved. The defect is closed in the operating room, catheterization laboratory or hybrid suite. The closure is directly through the right ventricle (RV) free wall, under echocardiographic guidance while the heart is beating. The visualization of the defect by echocardiography is better than direct visualization when the patient is on CPB.

The procedure requires mini-sternotomy, opening of the pericardial sac and a purse string over the RV free wall. Under TEE guidance, an angio-catheter is inserted into the RV; a wire is advanced through the catheter and maneuvered through the VSD into the left ventricle. An appropriate-sized deliver sheath is introduced over the wire through the RV free wall and the VSD into the left ventricle. The device is then placed in a fashion similar to the catheterization procedure.

The next frontier in tackling the VSD will be to do the procedure without sternotomy under robotic guidance. In addition, advances in development of delivery sheaths that can be maneuvered easily through the VSD during perventricular closure, improvement in device designs that may prevent rhythm issues and impingement of the cardiac valves will add significantly to optimal outcomes for the patient.

Although we have been able to close perimembranous VSD with perventricular technique in the past, we have stopped using devices in the perimembranous position because of high risk complete heart block. Once devices that will obviate this issue are available, perventricular approach will become an attractive option for perimembranous VSD as well.

In summary, device closure of VSD has come a long way with different options available as to the delivery of devices. We, however, can and will go further in improving the minimally invasive ways of device delivery, improving in design of devices, developing better delivery catheters and researching on robotic procedures.

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Abstract Title: Evolving Concepts in the Management of Arrhythmias in Adults with Congenital Heart Disease
Presenter: Anjan S Batra, MD; Director of Electrophysiology; Children's Hospital of Orange County; Associate Professor of Clinical Pediatrics; University of California, Irvine; Irvine, CA USA

Objective

- Understand the scope of the problem of arrhythmias in adults with congenital heart disease (CHD).
- Have an understanding of the arrhythmias encountered in common forms of CHD.
- Risk stratify these patients for sudden death, ICDs, catheter ablation and surgical therapy.
- Understand the unique approaches of treating arrhythmias in adults with CHD.

Abstract

Adults with congenital heart disease are a rapidly growing segment of the population. There are more adults than children living with CHD.[1] The incidence of arrhythmias increases with age and, by adulthood, arrhythmias are the leading cause of hospital admissions. Sudden death of presumed arrhythmic etiology is the most common cause of mortality. Criteria for risk stratification for primary and secondary prevention of arrhythmias and sudden death continue to evolve.[2] Other evolving concepts in the management of arrhythmias in adults with congenital heart disease include newer medications (Ibutelide, Dofetilide),[3] advances in 3D Electro-anatomical mapping,[4] and newer catheter technology (cooled tip chili catheter). Advances in pacing include newer devices capable of antitachycardia pacing,[5] resynchronization,[6] and defibrillation. Ablation of arrhythmia substrates can be done in the catheterization laboratory and surgically in the operating room.[7] Other hybrid approaches are also being developed to address these complex arrhythmias.[8] Despite huge improvements in the recent past, available therapies for treatment of arrhythmias in adults with congenital heart disease remain inadequate.

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Abstract Title: **Pediatric Cardiac Transplantation: The Next 25 Years**
Presenter: **Daniel Bernstein, MD; Chief, Division of Pediatric Cardiology; Lucille Packard Children's Hospital; Co-Director, Children's Heart Center; Alfred Woodley Salter and Mabel G. Salter Endowment; Professor of Pediatrics; Stanford University Medical Center; Palo Alto, CA USA**

Objective

1. Improve competence in managing difficult heart transplant issues leading to improved survival and reduced morbidity.
2. Increase understanding of new mechanisms for managing high risk patients with heart failure to improve pre and post transplant outcomes.

Abstract

Cardiac transplantation has evolved significantly over the past quarter century, moving from a highly experimental procedure to one that has become routine, complete with standardized protocols and a successful multi-center registry. Over that time frame, one-year survival has increased from 75% to over 90% and there are an increasing number of true long-term survivors. The next era of transplant medicine will be one of increasing innovation, with the development of new technologies, translation of molecular discoveries to the bedside, and the application of the power of personalized medicine through discoveries in genomics and pharmacogenomics. We will review several challenges for the future:

- pediatric mechanical circulatory support including the development of smaller and less traumatic and risky pediatric support systems;
- the non-invasive diagnosis of rejection using molecular tools for analyzing genome-wide immune pathway activation,
- transplantation of the highly sensitized patient, representing an increasing population of pediatric heart transplant candidates, and
- pharmacogenomics applied to pediatric heart transplantation, allowing clinicians to individualize immunosuppressive regimens.

Through these innovations, the next era of pediatric heart transplantation will be marked by a further reduction in mortality and morbidity and an extension of the benefits of transplantation to patients who would have previously been ineligible.

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Abstract Title: Kawasaki Disease Update
Presenter: Jane C. Burns MD; Professor and Chief, Division of Allergy, Immunology and Rheumatology; Director, Kawasaki Disease Research Center; Rady CHildren's Hospital - Sand Diego; Department of Pediatrics; University of California, San Diego, School of Medicine; San Diego, CA USA

Objective

1. To update current knowledge of epidemiology and treatment of Kawasaki disease (KD)
2. To review cardiac outcomes and controversy on appropriate follow-up of KD patients
3. To introduce AHA risk stratification scheme
4. To describe range of potential cardiovascular sequelae in adulthood

Abstract

Kawasaki Disease (KD) is the leading cause of acquired heart disease in children in the developed world.[1] Typically, KD presents in children under the age of five years as a febrile illness with mucocutaneous changes. A subset of patients will develop permanent damage to the arterial wall, valve leaflets, and myocardium. The acute phase of the illness is self -limited and the diagnosis may be missed. If untreated, KD can result in coronary aneurysms in 25% of patients.[2] Patients who suffer coronary artery damage may develop thrombosis or stenotic lesions associated with the aneurysms and are at risk of myocardial infarction, sudden death, and congestive heart failure.[3,4] The prognosis for adults who recovered from KD without coronary aneurysms is postulated to be good, but longitudinal studies have not been performed to test this hypothesis.

In 2004, the American Heart Association issued updated guidelines for the care and management of children with KD.[5] However, studies of the optimal therapy and management of the sequelae of KD in children and adults have not been performed. It is estimated that over 4,000 new cases of KD are diagnosed in the U.S. each year. In Japan, where the incidence is approximately 10-fold higher as compared to the U.S., more than 12,000 new cases are diagnosed each year. Seasonality of cases, nationwide epidemics, and the self-limited nature of the acute illness suggest an infectious trigger but no causative agent has been identified. Genetic influences on disease susceptibility and outcome have been identified and the current paradigm proposes that KD results from exposure to a common agent that triggers the syndrome only in genetically susceptible hosts.

The self-limited clinical syndrome is recognized through a constellation of clinical signs that include: fever for at least 4 days associated with rash, conjunctival injection, erythema of the lips and oropharynx, edema of the hands and feet, erythema of the palms and soles, and, in the convalescent phase, periungual desquamation. Up to 25% of untreated children will develop permanent damage to the coronary arteries with inflammatory cell infiltration of the arterial wall, destruction of the internal elastic lamina, necrosis of smooth muscle cells, myointimal proliferation,

and subsequent aneurysm formation. Aneurysms of systemic, extra-parenchymal muscular arteries also occur in a subset of patients with coronary aneurysms. The proximal coronary arteries can be readily imaged in infants and children using transthoracic echocardiography that permits reliable and reproducible measurement of the internal diameter of the proximal right and left anterior descending coronary arteries (RCA and LAD) and the expression of these measurements as standard deviation units (Z score) normalized for body surface area (BSA).

Administration of a single dose of intravenous immunoglobulin (IVIG) in conjunction with aspirin within the first ten days after fever onset reduces the incidence of aneurysms from 25% to 3-5%. The AHA guidelines have divided these patients into five groups based on coronary artery Z scores and morphology of the coronary artery lesions. Approximately 30% of IVIG-treated children with KD will develop transient dilatation of the coronary arteries (Z score ≥ 2.5 for the RCA or LAD, AHA Risk level II). Another 5-10% will develop coronary artery aneurysms, which in some cases can be attributed to delayed diagnosis and treatment.

Management of the KD patient with evolving aneurysms should focus on halting the inflammatory process, decreasing myocardial oxygen demand, and preventing thrombosis. Use of anti-inflammatory agents to block TNF alpha (infliximab) and to downregulate T-cell activation (cyclosporine, tacrolimus) are currently being studied to decrease inflammation. Beta-blockade and transfusion may have a cardioprotectant role. Anti-platelet therapy and systemic anti-coagulation have roles in prevention of thrombosis.

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Abstract Title: Assessment of the Hybrid Approach to the Management of HLHS
Presenter: John P. Cheatham, MD, FAAP, FACC; FSCAI; George H. Dunlap Endowment Chair in Interventional Cardiology; Director, Cardiac Catheterization and Interventional Therapy; Co-Director, The Heart Center, Nationwide Children's Hospital; Professor, Pediatrics and Internal Medicine, Cardiology Division; The Ohio State University College of Medicine; Columbus, OH USA

Objectives

- Understand the techniques for performing Hybrid Stage I Palliation
- Describe the indications & contraindications of the Hybrid procedure
- List the potential advantages & disadvantages of the Hybrid approach.

Abstract

Hypoplastic Left Heart Syndrome (HLHS) is uniformly fatal with approximately 90% of patients dying within the first month of life without some sort of intervention. The objective of Stage I palliation for single ventricle physiology is to provide an unobstructed systemic outflow tract, unrestrictive interatrial communication, a controlled source of pulmonary blood flow, and a reliable source of coronary blood flow. The

Norwood procedure described in 1981, cardiac transplantation described in 1986, and the Sano modification of the Norwood procedure in 2001, all met the above objectives. However, while individual centers worldwide have had success with one or more of the surgical strategies, the learning curve has been steep and neurodevelopmental outcomes questionable.

In 1999 while at Nemours Cardiac Center in Orlando, a "comprehensive" Hybrid approach was initiated, where surgical and transcatheter therapies were merged in order to flank one comprehensive open heart surgical procedure with two off CPB procedures: initially Hybrid Stage I palliation and a final transcatheter Fontan completion. From 2002 to present, members of The Heart Center at Nationwide Children's Hospital have promoted this approach as not only an acceptable alternative to "high risk patients", but also to all patients with HLHS, complex single and two-ventricle anatomy, who are being considered for conventional surgical procedures or cardiac transplantation.

Hybrid Stage I Palliation evolved into placing surgical RPA and LPA bands using Gore-Tex strips through a small median sternotomy off CPB, followed by a PDA stent delivered through a sheath placed in the MPA through a small purse-string suture using fluoroscopic guidance. Finally, creation of an adequate ASD is performed electively prior to discharge using a BAS catheter. In the case of severely restrictive ASD or intact atrial septum, the communication is created first by several different techniques, including: RF perforation, stent implant, or static balloon septoplasty. This palliation meets the criteria of 1) control & protects PBF, 2) provide reliable systemic cardiac output, and 3) create unobstructed flow from the LA.

Both Giessen University in Germany and NCH in Columbus championed this approach and published extensively so others could follow… from the initial techniques, to lessons learned, to intermediate results and follow-up. Using this Hybrid approach, contraindications to therapy in these sick neonates virtually disappeared. Size of patient, size of ascending aorta, poor RV function with significant TR, and avoidance of blood products were no longer a contraindication for palliation…unlike for the Norwood or Sano surgical procedures. Eventual biventricular repair or later cardiac transplant strategies could also benefit from Hybrid Stage I palliation. Only retrograde aortic arch flow obstruction seemed to be a contraindication to this novel approach.

In 2008, Galantowicz, et al published NHC's intermediate results in The Annals of Thoracic Surgery. From July 2002 – June 2008, 80 neonates underwent PA bands and PDA stent weighing 1.1 – 4.0 Kg, average age of 5 days, with 65 HLHS, 13 complex SV, and 2 complex 2V. There were 55 patients with typical HLHS and uniform risks who were analyzed. Procedural mortality was 2%, interstage mortality 6%, interstage reinterventions 26%, Comprehensive Stage II repair mortality 6% with no open sternum or ECMO support needed. The overall cumulative survival was 87% (92% for "usual risks"), which was virtually identical to that reported by Giessen as well as quite favorable to the contemporary reports from Boston and Philadelphia.

A concern with the Hybrid approach is the persistence of requiring retrograde aortic arch flow for the additional 4-6 months until Comprehensive Stage II is performed, as well as the challenges of PDA stent and PA band surgical removal. Thus far, interstage retrograde aortic flow obstruction occurs in ~ 20% of patients and can be successfully treated with percutaneous stent therapy. All PDA stents have been able to be removed surgically. However, further studies are underway to assess the neurodevelopmental outcomes of these patients, with correlation with transcranial Doppler and MR imaging… both through an NIH grant and a multi-institutional study.

While some centers have only offered the Hybrid approach to those high risk neonates who are not suitable candidates for the Norwood or Sano repairs, the results thus far suggest that the Hybrid approach should be an evenly discussed option with all patients, regardless of their risk scale. Improvement in both mortality and neurodevelopmental outcomes must override conventional strategies and thinking.

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

CONGENITAL CARDIOLOGY TODAY

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Brief Statement for:
Melody® Transcatheter Pulmonary Valve
Ensemble® Transcatheter Valve
Delivery System

The Melody TPV is indicated for use as an adjunct to surgery in the management of pediatric and adult patients with the following clinical conditions:

- Existence of a full (circumferential) RVOT conduit that was equal to or greater than 16 mm in diameter when originally implanted and
- Dysfunctional RVOT conduits with a clinical indication for intervention, and either:
  -regurgitation: ≥ moderate regurgitation, or
  -stenosis: mean RVOT gradient ≥ 35 mm Hg

Contraindications: None known.

Warnings/Precautions/Side Effects:

- **DO NOT implant in the aortic or mitral position. Preclinical bench testing of the Melody valve function and durability will be extremely limited when used in these locations.**
- DO NOT use if patient's anatomy precludes introduction of the valve, if the venous anatomy cannot accommodate a 22-Fr size introducer, or if there is significant obstruction of the central veins.
- DO NOT use if there are clinical or biological signs of infection including active endocarditis.
- Assessment of the coronary artery anatomy for the risk of coronary artery compression should be performed in all patients prior to deployment of the TPV.
- To minimize the risk of conduit rupture, do not use a balloon with a diameter greater than 110% of the nominal diameter (original implant size) of the conduit for pre-dilation of the intended site of deployment, or for deployment of the TPV.
- The potential for stent fracture should be considered in all patients who undergo TPV placement. Radiographic assessment of the stent with chest radiography or fluoroscopy should be included in the routine postoperative evaluation of patients who receive a TPV.
- If a stent fracture is detected, continued monitoring of the stent should be performed in conjunction with clinically appropriate hemodynamic assessment. In patients with stent fracture and significant associated RVOT obstruction or regurgitation, reintervention should be considered in accordance with usual clinical practice.

Potential procedural complications that may result from implantation of the Melody device include the following: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, pain at the catheterization site.

Potential device-related adverse events that may occur following device implantation include the following: stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, hemolysis.

For additional information, please refer to the Instructions for Use provided with the product.

CAUTION: Federal law (USA) restricts this device to sale by or on the order of a physician.

Hope,
Restored.

Medtronic

Melody® TPV Receives FDA HDE Approval

Melody® Transcatheter Pulmonary Valve (TPV) Therapy: A revolutionary, non-surgical option for children and adults with RVOT conduit dysfunction.

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Humanitarian Device.
Authorized by Federal law (USA) for use in pediatric and adult patients with a regurgitant or stenotic Right Ventricular Outflow Tract (RVOT) conduit (≥ 16 mm in diameter when originally implanted). The effectiveness of this device for this use has not been demonstrated.

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