CONGENITAL CARDIOLOGY TODAY

Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

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International Edition

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Sep. 15-19, 2014; Chicago, IL USA
www2.aap.org/sections/cardiology/pediatric_cardiology/2014/

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Melody Valve Implantation Using a Double-Balloon "Flower-Blossom" Technique

By Carolyn Wilhelm, MD; Jason Swinning, RCIS; Matt Sisk, RCIS; Ralf Holzer, MD, MSc,

There are videos associated with figures 1, 2, 3, 5, 6, 7, 8, 9 and 10 in this article.

You may view these videos simply by clicking the web link in the appropriate figures.

"Melody Valve implantation is an established procedure. However, placing the valve in patients with very short conduits can be challenging."

Background

Melody Valve implantation is an established procedure. However, placing the valve in patients with very short conduits can be challenging. While folding of the Melody valve stent has been described by Jounes Boudjemine when a shorter valve stent is required,[1] this may not necessarily be appropriate in patients who also have somewhat small branch pulmonary arteries and distal conduit stentosis, rendering the use of a single balloon as a less suitable technique. Placing a stent on two balloons for a distal conduit stenosis was originally described by Oliver Stumper.[2] In this report we describe the first case of a Melody Valve being implanted using this technique over two balloons simultaneously placed in the right and left pulmonary arteries.

Methods

A 17-year-old female patient with a history of Tetralogy of Fallot repair at the age of eight months, subsequently required bilateral proximal pulmonary artery stent placement using two 25mm Genesis XD stents (Cordis, Bridgewater, NJ - www.cordis.com) at the age of six years. At the age of 15 years, she underwent placement of a 20mm valved Core Matrix RVPA conduit and intraoperative placement of an additional 19mm Genesis XD (Cordis, Bridgewater, NJ) stent to the LPA, balloon angioplasty of both pulmonary artery stents (to 10mm), as well as resection of any protruding stent meshwork. On echocardiography follow-up, she developed dehiscence of a valve leaflet, prolapsing into the RVOT with free pulmonary insufficiency, as well as evidence of some narrowing across the distal end of the RVPA conduit, with a systolic

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Doppler gradient of about 50mmHg, and mild RA and RV enlargement. She was asymptomatic with a fairly sedentary lifestyle and a cochlear implant for partial deafness. ECG documented SR with RBBB and a QRS duration of 126ms. An MRI could not be performed due to the cochlear implant and a limited CT scan due to motion artifact, documented narrowing of the conduit at its distal end (8*14mm), slightly small branch PA stent diameters (7-9mm), and mild RA and RV enlargement with normal RV function. She was taken to the catheterization laboratory with the intent of rehabilitating any conduit and branch pulmonary artery stenosis, followed by Melody Valve implantation.

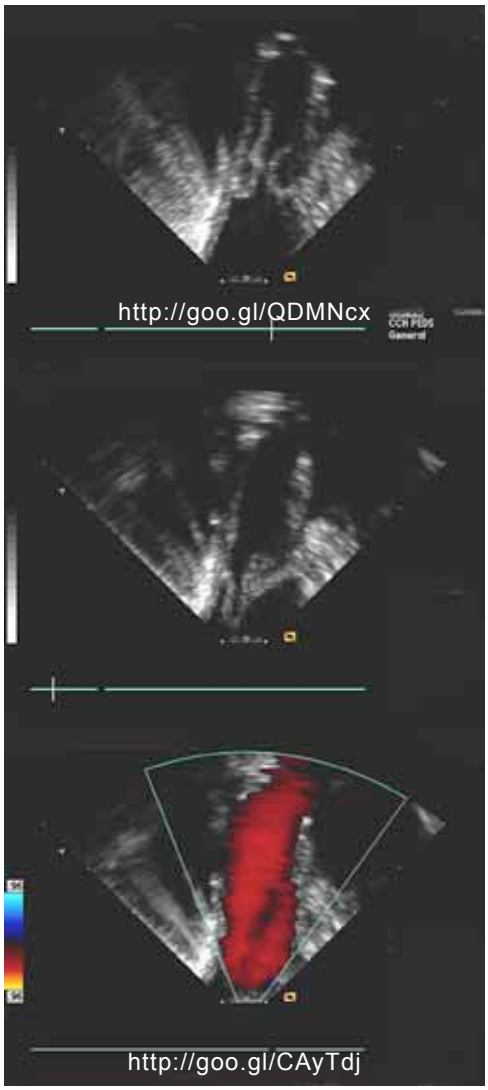

Figure 1. Baseline intracardiac echocardiography. Top image: Leaflets of the Core Matrix conduit opening into the conduit (systole). Middle image: The leaflets prolapsing into the right ventricular outflow tract. Bottom image: Free pulmonary insufficiency.

The procedure was performed under general anesthesia. An 18Fr short sheath was placed inside the right femoral vein and a 6Fr sheath in the right femoral artery. Baseline right and left heart catheterization was performed, documenting no significant gradient through the RPA and LPA stents, but a 25mmHg gradient across the distal conduit, as well as a 10-15mmHg gradient at the Core Matrix valvar annulus. The RV pressure was 60% systemic. Intracardiac echocardiography documented flail Core Matrix conduit leaflets prolapsing into the RVOT (Figure 1).

Figure 2. 3D reconstruction of a rotational angiography performed under rapid RV pacing, documenting a distal conduit stenosis and some size discrepancy of the proximal PA stents when compared to the distal vessels.

A rotational angiography was obtained inside the distal conduit under rapid RV pacing, with 3D reconstruction documenting some narrowing of the distal conduit and the proximal stented branch PAs being somewhat smaller than the distal vessels (Figure 2).

This was followed by 2-dimensional angiography, documenting, again, the narrowing of the distal conduit (Figure 3), measuring distally 12.1mm, at the RVOT insertion 16.6mm, with the length from RVOT insertion to the branch PA stent edges being ~22-23mm.

Further angiographies were obtained in the LPA as well as RPA, followed by balloon angioplasty of both in situ stents, using 12mm*2cm Atlas Gold balloons (Bard Peripheral Vascular, Tempe, AZ - www.bardpv.com) (Figure 4).

This was then followed by coronary evaluation during simultaneous inflation of two 12mm*4cm ZMedII balloons (BBraun, Bethlehem, PA - www.bisusa.org) in RPA and LPA with a combined effective diameter of 19.6mm, showing no evidence of any coronary compression (Figure 5).

At low inflation pressures of 2 atm, the two balloons had a combined waist of just

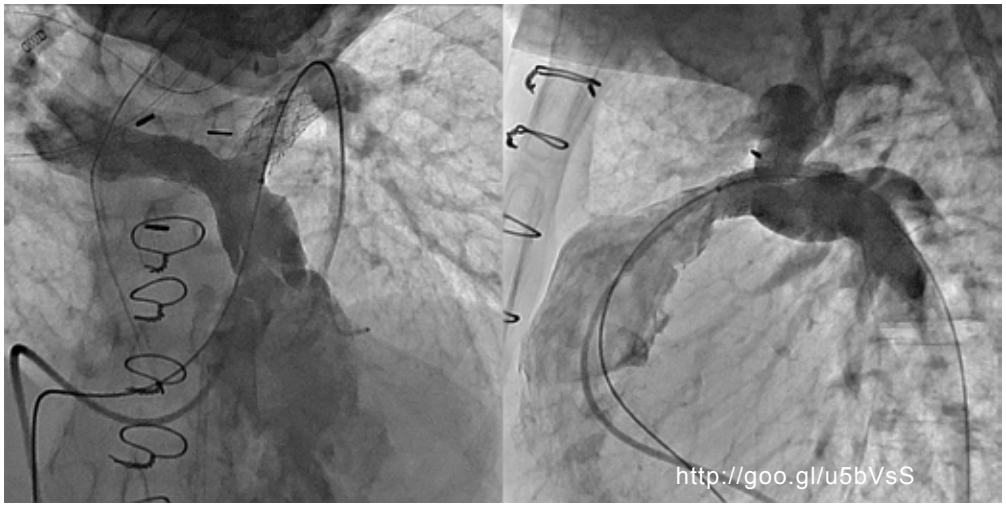

Figure 3. Baseline angiography documenting narrowing at the distal end of the Core Matrix conduit (proximal to the branch PA stents), as well as the flail Core Matrix valve leaflet.

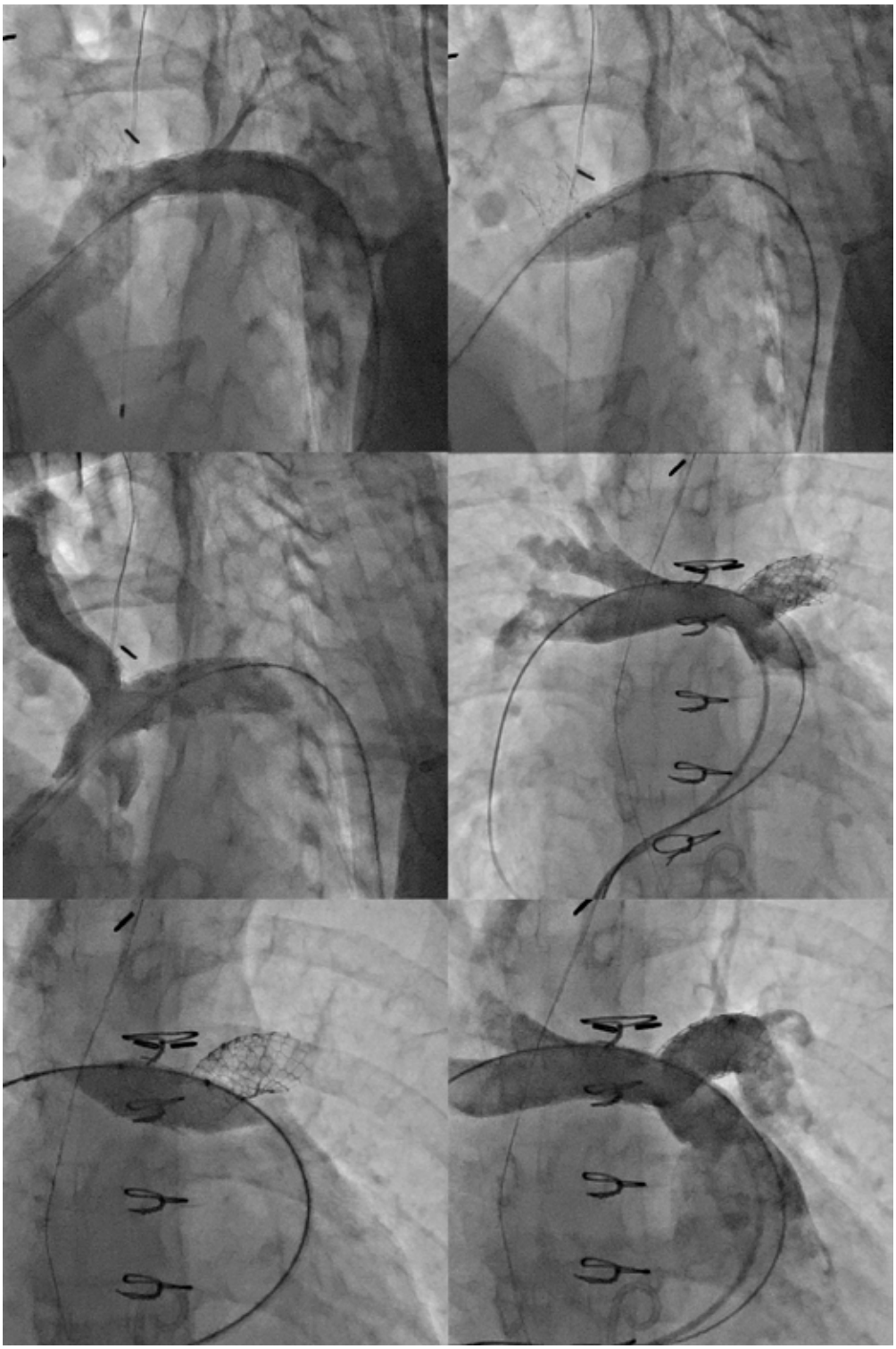

Figure 4. Angioplasty of both PA stents. The top left: Some instent stenosis of the LPA stent, as well as a larger distal LPA diameter. Top right: LPA balloon angioplasty. Middle left: Angiography post-LPA angioplasty (improved LPA appearance, as well as RPA stenosis). Middle right: RPA angioplasty with some proximal size discrepancy. Bottom right: RPA angioplasty. Bottom left: Angiography post RPA angioplasty (improved RPA appearance).

12.3mm. It was, therefore, decided to change to two 10mm*4cm ZMedII balloons (BBraun, Bethlehem, PA) with a combined diameter about 16.4mm and perform double-balloon angioplasty with inflation pressures of up to 12 atm, followed subsequently (after an unremarkable angiography) by double-balloon angioplasty using two 12mm*4cm ZMedII balloons (BBraun, Bethlehem, PA) with inflation pressures of up to 12 atm and only a trivial residual waist. The angiography documented residual mild narrowing at the distal conduit (13.1mm), in addition to pre-existing pulmonary insufficiency (Figure 6). It was therefore, decided to perform stent implantation across the RVPA conduit. To evaluate if and how a single 20mm balloon could be stabilized within the distal conduit given its close proximity to the branch PA stents, balloon angioplasty was performed using a 20mm*3cm ZMedII balloon (BBraun, Bethlehem, PA), and it was immediately apparent that stent implantation using this modality would not be feasible, with the balloon milking closer to the RVOT and not capturing the distal conduit (Figure 7).

Given the need to treat the distal end of the conduit, the short length of the conduit of just 22-23mm, and the fact that a 3110 Palmaz XL stent would shorten to just over 26mm when expanded to 20mm, it was felt that a "flower-blossom" technique as originally described by Oliver Stumper[2] should be utilized. For this purpose, after placing a 16Fr long sheath within the distal conduit using an ultra-stiff wire positioned in the RPA, a second stiff wire was positioned through the sheath into the LPA. A 3110 Palmaz XL stent (Cordis, Bridgewater, NJ) was mounted over two 12mm*4cm ZMedII balloons, to allow the balloons to be slightly longer than the stent itself to facilitate simultaneous advancement to the RPA and LPA. The stent-double-balloon assembly was advanced over the two wires and once positioned far enough overlapping the RPA and LPA stents, the balloons were inflated simultaneously, nicely molding the Palmaz XL stent to the RPA and LPA (Figure 8).

Further balloon angioplasty was performed on the distal stent using two high-pressure 12mm*2cm Atlas Gold balloons, with a subsequent angiography documenting excellent stent positioning (Figure 8). At this point preparations were made for transcatheter Melody Valve implantation (Medtronic, Minneapolis, MN - www.medtronic.com/melody/), and given the inability to expand a single larger balloon distally enough (without milking proximally) due to interference of the branch PA stents, it was felt that the Melody Valve should be implanted using the same flower-blossom technique. After readily inspecting and prepping the valve with standard technique, the valve was mounted over two 12mm*4cm ZMedII balloons. To evaluate what sheath would be required for valve implantation, a 20Fr sheath was cut, and while the mounted valve could be advanced through the cut-off sheath, the pushability was rather tight, and it was therefore, decided to use a 22Fr long sheath for valve implantation. Initially, attempts were made to advance the sheath over a single ultra-stiff wire positioned in the RPA, but due to the stiffness of the

Melody® Transcatheter Pulmonary Valve Ensemble® Transcatheter Valve Delivery System

Indications: The Melody TPV is indicated for use in a dysfunctional Right Ventricular outflow Tract (RVOT) conduit (≥16mm in diameter when originally implanted) that is either regurgitant (≥ moderate) or stenotic (mean RVOT gradient ≥ 35 mm Hg)

Contraindications: None known.

Warnings/Precautions/Side Effects:

- DO NOT implant in the aortic or mitral position.
- 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: 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, and pain at the catheterization site.

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

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

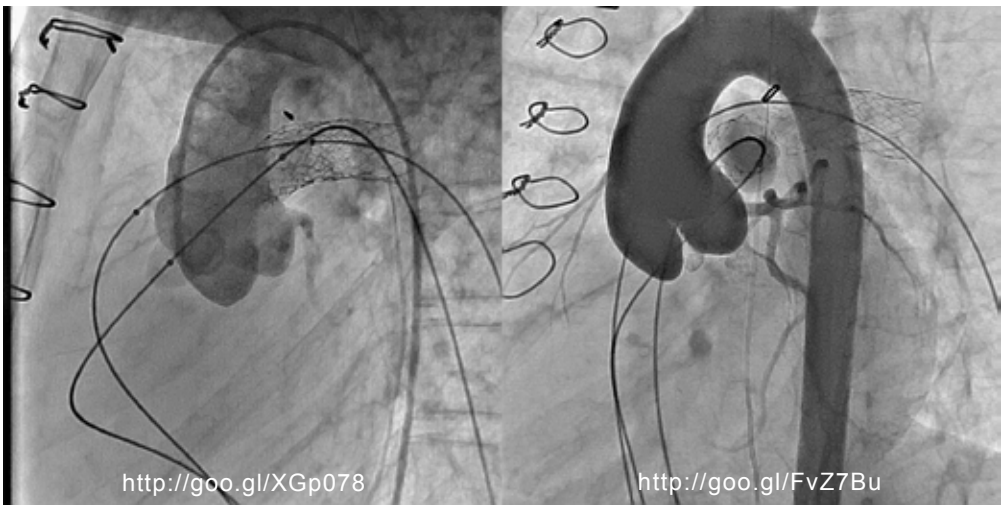

Figure 5. Coronary evaluation (aortogram) during double-balloon inflation in RPA and LPA (no evidence of coronary compression).

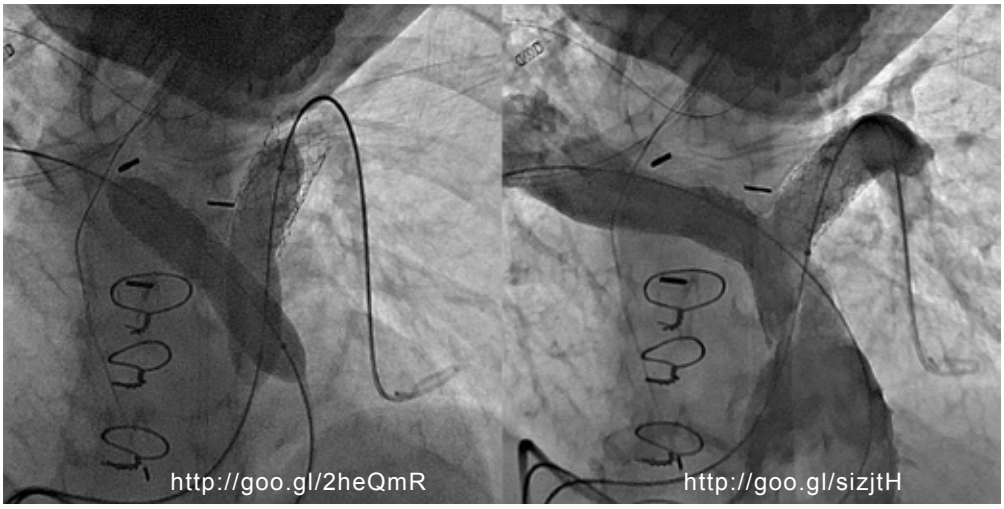

*Figure 6. Left: Double balloon angioplasty of the Core Matrix conduit using two 12mm*4cm ZMedII balloons. Right: Angiography post angioplasty documenting residual narrowing of the distal conduit, as well as free pulmonary insufficiency.*

sheath, those attempts were unsuccessful. Therefore, the wire was repositioned to the LPA, which had been the more difficult to engage vessel earlier in the procedure, and after prolonged attempts the sheath was eventually positioned within the distal third of the Palmaz XL stent. At this point a second ultra-stiff wire was positioned to the RPA and the mounted Melody-double-balloon assembly advanced over the two wires across the sheath. Some kinking of the sheath required gentle retraction, and while the assembly could be moved half way into the Palmaz XL stent, we were unable to easily position it more distally. It was clear at this point that the long sheath, as well as the stiff wires, were hampering the attempts to advance the assembly more distally, and eventually a combination of sheath retraction to the RA, exchanging the ultra-stiff RPA wire to a softer 035" angled glide wire, and pulling back the LPA wire to the proximal LPA, allowed the assembly to be freed from the stent and by buckling the balloon within the RA, the assembly eventually was positioned in the desired position with each of the two balloons engaging one of the branch PAs (Figure 9).

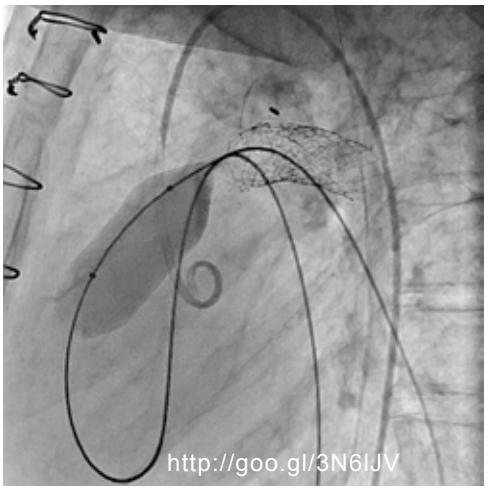

*Figure 7. Test angioplasty of the Core Matrix conduit using a single 20mm*3cm ZMedII balloon with inflation initially commenced with the balloon positioned closely to the RPA stent, resulting in the balloon milking into the RVOT in a position unsuitable for appropriate Melody Valve placement.*

At this point the valve was expanded, followed by double-balloon angioplasty using a 14mm*4cm ZMedII balloon to the RPA (given that the RPA stent still appeared slightly small) and a 12mm*4cm ZMedII balloon to the LPA. To provide a more round appearance of the valve, a 20mm*4cm ZMedII balloon was then advanced into the Melody Valve and inflated towards the RPA with the shoulder of the balloon initially positioned directly at the RPA stent origin. Again, during inflation it was clear that the balloon milked somewhat back towards the RVOT, further underlining the importance of not placing the Melody Valve on a single balloon in this patient (Figure 9).

A second set of hemodynamic data was obtained, documenting only a 2mmHg gradient through the RPA stent, and a 3mmHg gradient from distal conduit to RV, with systolic RV pressures being 27mmHg and systolic aortic pressures being 93mmHg (RV = 29% systemic). A final angiography documented no significant pulmonary insufficiency (Figure 9), and a final evaluation by intracardiac echocardiography documented the previously seen flail Core Matrix leaflet being trapped behind the stents with unrestricted mobility of the Melody Valve leaflets and only trace pulmonary

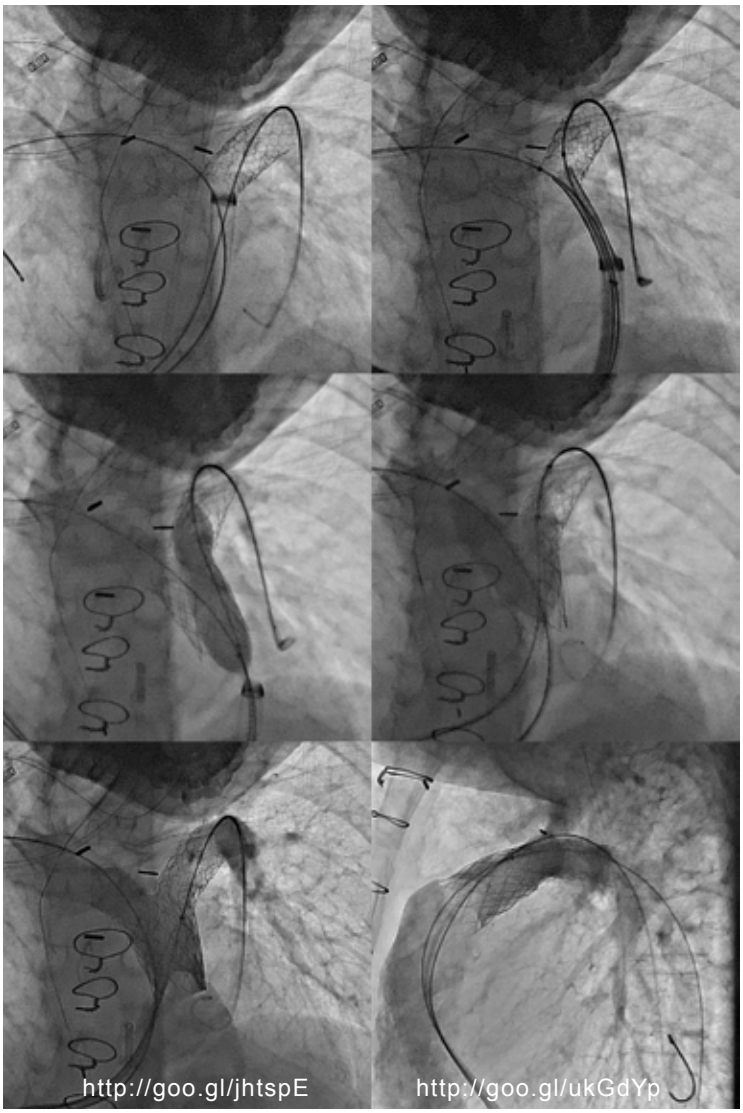

Figure 8. Implantation of a 3110 Palmaz XL stent into the Core Matrix conduit using a "flower-blossom" technique. Top left: A 16Fr long sheath positioned over two stiff wires into the distal conduit. Top right: Positioning of the 3110 Palmaz XL within the distal conduit stent mounted on two 12mm 4cm ZMedII balloons. Middle left: Balloon expansion of the stent folding it into the RPA and LPA simultaneously (The RPA balloon did not have sufficient contrast to appreciate its contour more clearly). Middle right: Double-balloon angioplasty of the stent using two high-pressure 12mm2cm Atlas Gold balloons. Bottom left and right: Angiography after stent placement documenting good positioning with some "funneling" at the distal end on lateral projection.

insufficiency. The patient recovered well, and was discharged home the following day.

Conclusion

In a conclusion, Melody Valve implantation can be somewhat challenging in patients with unusual anatomy. While folding of the Melody valve stent is a possibility in some patients with a short RVPA conduit, it is important to also consider mounting the Melody Valve on two balloons as an alternative technique, especially when the size of the branch pulmonary arteries is a lot smaller than the intended diameter of the Melody Valve, and when the distal end of a short conduit needs to be captured. This is even more so the case in the presence of bilateral PA stents which easily lead to a single large balloon milking proximally. This was the first case describing implantation of a

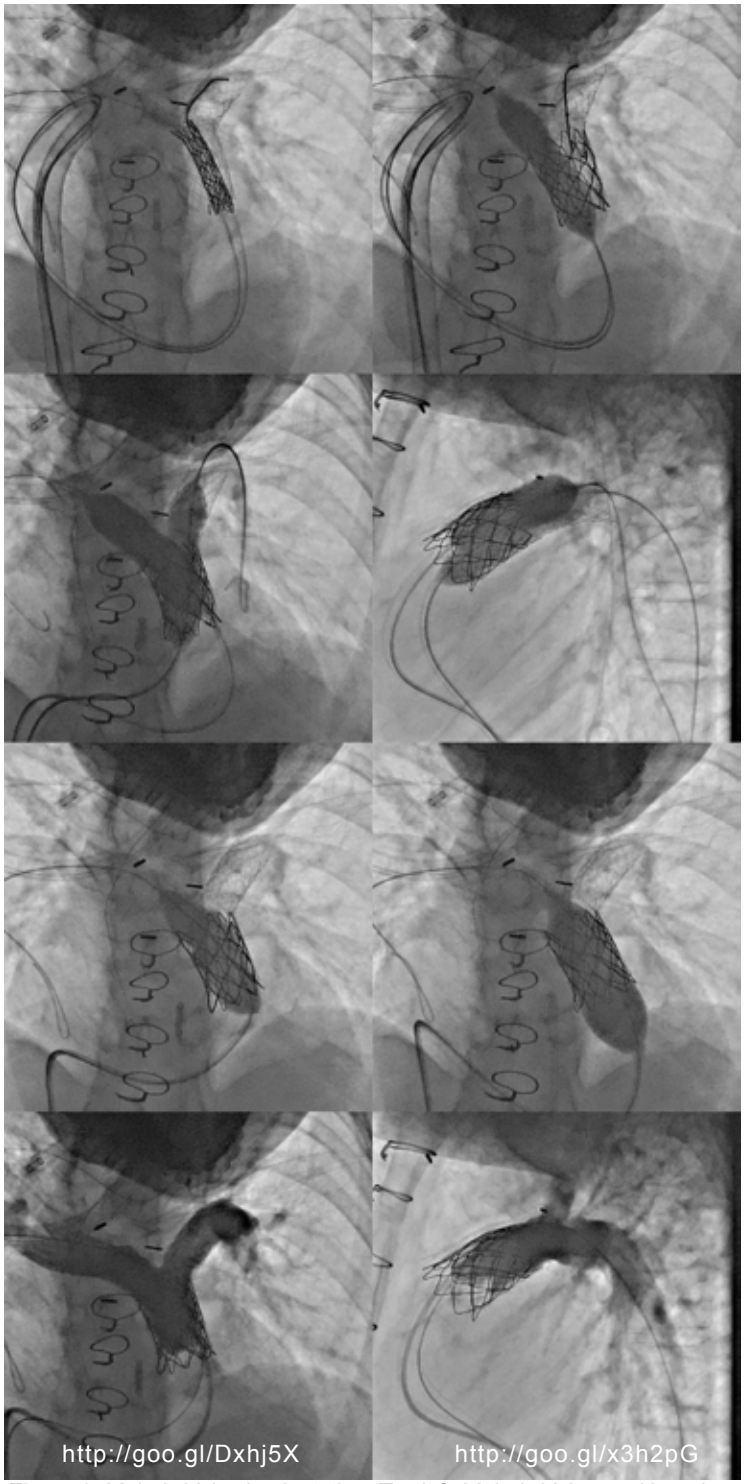

*Figure 9. Melody Valve implantation. Top left: Melody Valve positioned on two 12mm*4cm ZMedII balloons, initially positioned using a 22Fr long sheath (now in RA). The RPA wire had to be exchanged to an 035" angled glide wire and the LPA wire pulled back with the sheath retracted to the RA to allow the valve to be freed from the insitu XL stent and to be appropriately positioned distally. Top right: Expansion of the Melody valve on two 12mm*4cm ZMedII balloons – The LPA balloon unintentionally did not have any contrast and is only faintly visible (but the distal marker can be noted). Second row: Double balloon angioplasty of the Melody Valve using a 14mm*4cm ZMedII balloon to the RPA, and a 12mm*4cm ZMedII balloon to the LPA. Third row: Expansion of the Melody Valve using a single 20mm*3cm ZMedII balloon – note the positioning of the balloon at the beginning of the inflation (left) and at the end (right) clearly documenting that mounting the Melody on a single larger balloon would have led to the valve being implanted significantly within the RVOT, due to the limitations by the distal smaller diameter stents in the branch PAs. Bottom row: Angiography after Melody Valve implantation documenting no significant pulmonary insufficiency and no residual angiographic narrowing.*

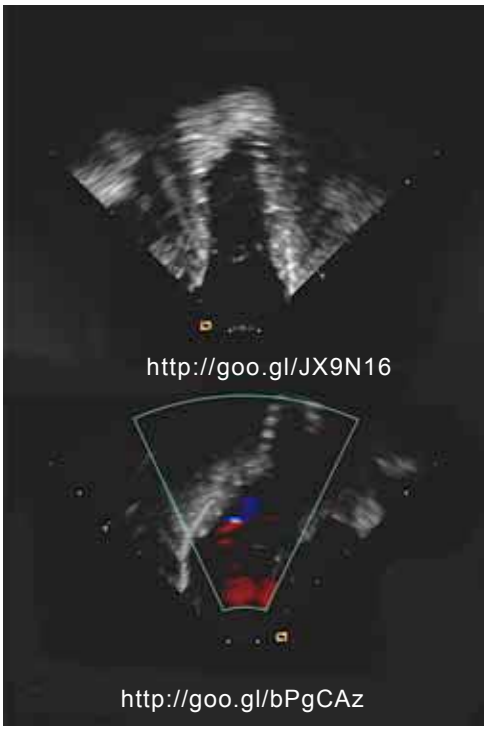

Figure 10. Intracardiac echocardiography at the end of the procedure, documenting the flail core matrix leaflet to be trapped beneath the stents, and normal movement of the Melody Valve leaflets, with only trace insufficiency.

Melody Valve using a "flower-blossom" technique over two balloons simultaneously, with the valve retaining appropriate competence after implantation.

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Letters to the Editor

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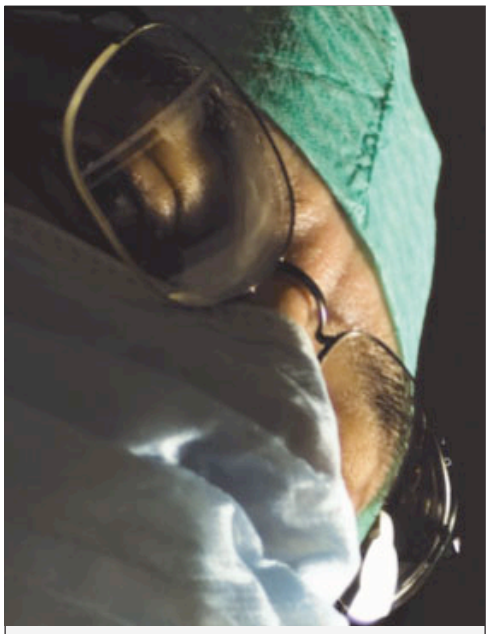
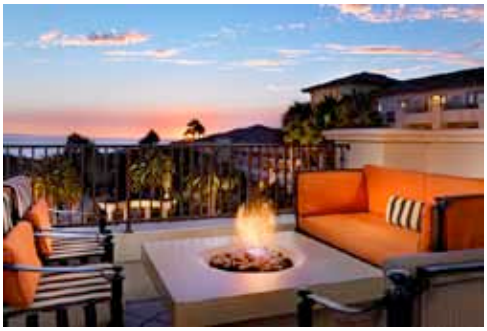


The 25th Annual Conference of the Western Society of Pediatric Cardiology

By Anjan S. Batra, MD; Mary E. Hickcox

The 25th Annual Conference of the Western Society of Pediatric Cardiology was hosted by Children's Hospital of Orange County (CHOC) at the beautiful St. Regis Monarch Beach Resort on April 11th-13th, 2014. Over 132 pediatric cardiologists attended the annual two and a half day society meeting.



St. Regis at Sunset, Dana Point, CA.

The conference focused on the controversy which exists with respect to the choice and timing of certain cardiac surgeries. Recognizing providers need to understand the advantages and disadvantages of a certain approach, multiple cases were reviewed, discussed and debated in order to recommend the best treatment for their patients.

This year's conference offered providers with up-to-date information about the following areas in order to ensure best outcomes for their patients: the latest advances in diagnostic techniques, new treatments for heart failure, cardiac screening of athletes, the risks of pregnancy in a patient with congenital heart disease, and the appropriate management of the asymptomatic patient with a positive genetic test for heart disease.

The academic nature of this conference allowed participants to interact during panel discussions, cases presented and debates. All were followed by a question and answer session. This format provided an opportunity to engage the physician learners, and enable them to interact on multiple levels. Conference attendees came primarily from California. About 30% of the audience came from the following West Coast Region: Alaska, Arizona, Colorado, Oregon, New Mexico, Washington State, Utah, and Texas.

Beginning at noon on Friday, April 11th, 2014, Dr. Paul Francis, current President of the Western Society of Pediatric Cardiology (WSOPC), opened the conference celebrating the 25th anniversary of the WSOPC. Dr. Francis discussed the history, current status and future plans of the WSOPC. In its genesis, the WSOPC was founded in 1960 as the California Society of Pediatric Cardiology. The society waned during the years prior to 1989 when Dr. Arno Hohn resurrected the society. In 2001, Dr. Norman Silverman effected a name change to the current Western Society of Pediatric Cardiology. Dr. Francis recapped the society's goals, advocacy efforts and topics planned for future annual conferences.

Throughout the conference, faculty took time to remember Dr. Arno R. Hohn, Professor of Pediatrics, Children's Hospital Los Angeles, University of Southern California, 1931-2014 and Dr. Beverly C. Morgan, Professor of Cardiology, University of California, Irvine, 1925-2014. During the Saturday evening Banquet and Abstract Awards, a time of memoriam was encouraged for members to honor the memory of Drs. Hohn and. Morgan.

Dr. Roberta Williams was the Keynote Speaker on Saturday morning, April 12th, 2014. Her talk covered "Bridging the Gap from Pediatrics to Adult Healthcare." This challenging topic exists as pediatric cardiology patients are the beneficiaries of early intervention, expert cardiac care and, as a result, are thriving into adulthood. Dr. Williams addressed how cardiologists can transition their pediatric patients through adolescence and into adult-centered health care. A *Transition Readiness Assessment Questionnaire* tool was included in the conference syllabus as an aid for cardiologists to implement into their practice.

Dr. Anjan Batra, MD (CHOC Children's) was the Program Chair for this year's conference. In collaboration with his colleagues, CHOC Planning Committee and CME Program Office, he developed a robust academic program. The agenda of the conference was divided into topic sessions.

Friday afternoon's session on ECHO was moderated by Dr. Mark Sklansky, (Children's Hospital of Los Angeles). Two case-based discussions on, "Where Does 3D Echo Add Value (Fetal, Transthoracic, Transesophageal)?" and "Invasive Fetal Cardiac Interventions: Future or Fad?" provided a platform for discussion from Brian Soriano, MD (Seattle Children's), Pierre Wong, MD (CHLA), Jay D. Pruetz, MD (USC/CHLA), Anita Moon-Grady, MD (UCSF) and Phil Moore, MD (UCSF).

Anjan S. Batra, MD; Roberta Williams, MD

"Throughout the conference, faculty took time to remember Dr. Arno R. Hohn, Professor of Pediatrics, Children's Hospital Los Angeles, University of Southern California, 1931-2014 and Dr. Beverly C. Morgan, Professor of Cardiology, University of California, Irvine, 1925-2014."

Friday's ECHO Session ended with a roundtable discussion on "Pediatric Echocardiography: Just for Pediatric Cardiologists?" This session reviewed the advantages and disadvantages of neonatologists performing echocardiograms, and was expertly addressed by Amir Ashrafi, MD (CHOC Children's), Nafiz Kiciman, MD (UCI), Anita Moon-Grady, MD (UCSF), Michael Puchalski, MD (Utah SOM) and Pierre Wong, MD (CHLA).

Friday ended with an Abstract Poster Reception in The Wine Room. Guests were able to review accepted abstract posters from 6 of the 13 abstracts submitted. The other 7 were accepted for an oral presentation on Saturday morning.

On Saturday morning, April 12th, 2014, the following oral abstract talks were presented:

- "Diagnostic Yield of Ambulatory ECG Patch Monitoring in Children from a National Registry" - Meena Bolourchi, MD (UCI)
- "Improving Screening Methods for Detection of Pediatric Hypertrophic Cardiomyopathy, 2014 Update" - Daniel Cortez, MD
- "Aortic Dilation in Eite United States Volleyball Players" - Christopher Davis, MD, PhD

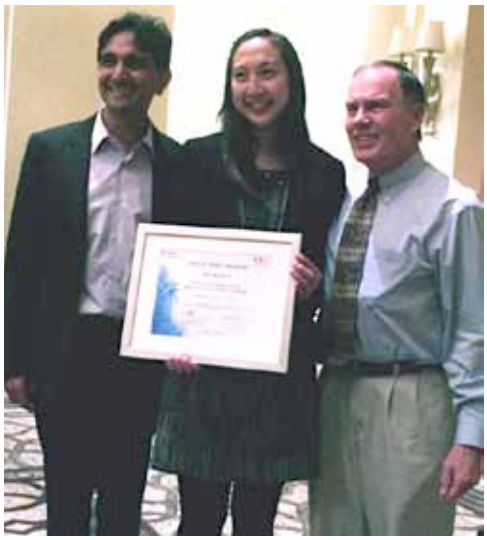
"Dr. Anjan Batra, MD (CHOC Children's) was the Program Chair for this year's conference.... The agenda of the conference was divided into topic sessions."

- "Deletion of ETS-1, a Gene in the Jacobsen Syndrome (11q-) Cardiac Critical Region, Causes Congenital Heart Defects Through a Cardiac Neural Crest Cell Migration Defect" - Paul Grossfeld, MD
- "Transcatheter Closure of Perimembranous Ventricular Septal Defect with the Amplatzer Vascular Plug II" - Nancy Hua, DO
- "Hemodynamic Effects of Withdrawal of Ventilatory Support in Intubated Children" - Arash Sabati, MD
- "Left Ventricular Tonic Contraction Frequently Precedes Dilation in Cardiomyopathy of Duchenne Muscular Dystrophy" - Jennifer Su, MD

A panel of judges in the audience graded the presentations on content, audience engagement, and staying within the time limits of the talk. All were excellent, and the grading was very close. The following individuals were awarded 1st, 2nd, and 3rd Place, and received a cash award and recognition at the evening's award banquet:

- **1st Place - Jennifer Su, MD (CHLA)** - "Left Ventricular Tonic Contraction Frequently Precedes Ddilation in Cardiomyopathy of Duchenne Muscular Dystrophy"
- **2nd Place - Meena Bolourchi, MD (UCI)** - "Diagnostic Yield of Ambulatory ECG Patch Monitoring in Children from a National Registry"
- **3rd Place - Christopher Davis, MD, PhD (RCHSD)** - "Aortic Dilation in Elite United States Volleyball Players"

In addition to her cash award, Dr. Jennifer Su was awarded the inaugural "Arno R. Hohn, MD Award for Best Abstract" in honor of Dr. Hohn. A professionally framed certificate was given to Dr. Su by Drs. Anjan S. Batra and Paul Francis.



Anjan S. Batra, MD; Joanna Su, MD; Paul Francis, MD.

After Dr. Williams keynote address, the first session was on Electrophysiology, moderated by Dr. Anjan Batra, (CHOC Children's). The first talk, "Management of the Genotype-Positive, Phenotype-Negative Child," was a case-based discussion with Yaniv Bar-Cohen, MD (USC/CHLA), Anjan S. Batra, MD (CHOC/UCI) and Kevin Shannon, MD (UCLA). They discussed ways to appropriately manage patients who are genotype positive and phenotype negative.

The next talk was a roundtable discussion: "Seattle Criteria for the Interpretation of ECG's in Athletes - Should These Become the New "Gold Standard?" Elizabeth V. Saarel, MD (Intermountain Healthcare, Utah), Anthony McCanta, MD (Children's Hospital Colorado) and Michael J. Silka, MD (CHLA) reviewed the Seattle criteria for interpretation of EKGs in athletes, and use of these criteria in the evaluation of athletes in the cardiologist's practice.

After lunch, a series of debates were moderated by Dr. Farhouch Berdjis (CHOC Children's) and Dr. Richard Gates (CHOC Children's) during the Surgery Session. The following debates were well-received, and generated audience interaction via audience response and Q&A:

- **The Sano/Norwood Procedure Leads to a More Favorable Outcome in Small Children Compared to the Hybrid Procedure** (Pro: Brian Reemtsen, MD (UCLA) and Con: Abraham Rothman, MD (Children's Heart Center, Nevada)) debated which surgical procedure(s) would be best suited for individual patients in practice. Additionally, each presented described the pros/cons of each commonly performed surgical procedure.
- **Transcatheter Therapy Should Replace Surgical Valve Replacement as Treatment of Choice for PS/PR** (Pro: Thomas Jones, MD (Seattle Children's) and Con: Tara Karamlou, MD, MS (UCSF)) reviewed pros/cons of implanting Melody Valves outside the current recommended age criteria.
- **Transcatheter PDA Closure in Preemies is the Procedure of Choice Versus Surgical Ligation** (Pro: Evan Zhan, MD (Cedars-Sinai) and Con: John Lamberti, MD (Rady Children's San Diego)) reviewed pros/cons of transcatheter PDA closure in preemies versus surgical ligation.
- **Stenting Should Never be the Procedure of Choice in Children with Pulmonary Artery Stenosis** (Pro: Frank Hanley, MD (Stanford) and Con: John W. Moore, MD (Rady Children's San Diego)) debated the factors which effect timing for intervention in patient with pulmonary stenosis. Also discussed were how to recognize and review patients who have the clinical indicators which necessitate intervention.
- **Coarctation: Surgery Should Always be the Treatment of Choice** (Pro: Richard Gates, MD (CHOC) and Con: Daniel Levi, MD (UCLA)) discussed the pros/cons of intervention versus surgical repair of coarctation.

The conference final day was on Sunday, April 13th, 2014. Dr. Anthony C. Chang, (CHOC Children's) moderated the first half of the Cardiac Intensive Care Session. Case Presentations of Heart Failure/ICU were discussed by a panel of experts: Stephen J. Roth, MD (Stanford) and Joanne Starr, MD (CHOC Children's). Where indicated, participants were encouraged to use the new modalities discussed in this session to manage heart failure. The panel discussed the new applications of old medicines in the ICU, and how to incorporate these practices into current treatment regimens.

The second half of the morning on Cardiac Intensive Care Session was moderated by

John W. Moore, MD; Farhouch Berdjis, MD; Richard Gates, MD; Frank Hanley, MD.

Jeannette Lin, MD; Priya Pillutla, MD; Roberta Williams, MD; Chuck Alejos, MD.

Juan "Chuck" Alejos, MD (Ronald Reagan UCLA Medical Center). Two roundtable discussions filled out the morning: "Cardiomyopathy, Stenotic Lesions, Single Ventricle: What is the Risk with Pregnancy?" was discussed by Jeannette Lin, MD (UCI), Priya Pillutla, MD (Harbor-UCLA) and Roberta Williams, MD (USC). They expressed ways to counsel parents and patients on the medical risks of pregnancy in patients with congenital heart disease. "No Boundaries - Mission Trips and CHD" was discussed by Juan C. Alejos, MD (UCLA), Michael Rebolledo, MD (CHOC) and Shaun P. Setty, MD (Miller Children's). They were able to describe opportunities, needs and barriers to providing cardiac care in Third World countries. The audience was encouraged to investigate and plan to participate in a medical outreach program in a Third World country.

The 26th Annual Conference of the Western Society of Pediatric Cardiology will be held March, 2015 in Aspen, Colorado, and will be sponsored by Children's Hospital, Colorado. Update information will be available at: www.wsopc.org.

CCT

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- Optionally, a picture of the author(s) may be submitted.
- No abstract should be submitted.
- The main text of the article should be written in informal style using correct English. The final manuscript may be between 400-4,000 words, and contain pictures, graphs, charts and tables. Accepted manuscripts will be published within 1-3 months of receipt. Abbreviations which are commonplace in pediatric cardiology or in the lay literature may be used.
- Comprehensive references are not required. We recommend that you provide only the most important and relevant references using the standard format.
- Figures should be submitted separately as individual separate electronic files. Numbered figure captions should be included in the main Word file after the references. Captions should be brief.
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Highlights from The 24th Annual Adult Congenital Heart Disease Program - June 10th to 13th, 2014

By Gary Webb, MD

The *24th Annual Adult Congenital Heart Disease Program* was hosted by the Heart Institute at Cincinnati Children's Hospital Medical Center June 10th to 13th, 2014. This is the major North American meeting for adult congenital heart disease experts each year, and the meeting has also recently been held at Skamania Lodge in Washington State, and in Toronto, Canada.

Over 300 registrants came to Cincinnati from all over the world, including a distinguished program faculty of pediatric and adult cardiologists and surgeons, and approximately 60 nurses and nurse practitioners, all from myriad countries and continents. Most attendees had a career commitment to ACHD practice.

In order to accommodate more themes and more faculty, two main meeting rooms ran parallel sessions for the last three days of the conference, with each area emphasizing different teaching points. Topic-based and case-based presentations were also made, typically on the same subjects.

Special themes for the program included:

- ACHD anesthesia and critical care
- Exercise and congenital heart disease
- ACHD catheterization and intervention
- Consensus guidelines on the recognition and management of arrhythmias in ACHD patients
- ACHD and heart failure
- Pregnancy and heart disease

Smaller symposia were held on MRI and ACHD; and Neurodevelopmental and Psychosocial Outcomes in ACHD patients.

The case-based presentations were made by four teams including: Switzerland, the Netherlands, the Montréal Heart Institute, and Hershey Medical Center. The teams did a great job of presenting a mix of interesting cases, and making sure the audience understood the main learning points. The teams used audience response questions to keep everyone engaged in the presentations and spark conversation.

"Mr. Redlegs" didn't miss our celebration at the Reds-Dodgers baseball game.

The program opened June 10th with a special all-day symposium focused on transition medicine titled, "Transition from Pediatric to Adult Health Care: The Cincinnati Summit."

The symposium's goal was to present examples of successful transition programs. Presentations included faculty from: The Hospital for Sick Children in Toronto; University Hospital Zürich; the Sickle Cell Transition Program and the Adolescent and Adult Congenital Heart Program at Cincinnati Children's Hospital.

Approximately 150 people participated and all attendees saw concrete examples of how to incorporate and build-on the success of other programs in this challenging area where children with congenital heart defects are all too often lost to follow-up, despite the need for lifelong care.

A special Cardiac Care Associates Program focused on international challenges and opportunities, and included presentations by cardiac care associates from Ireland, New Zealand, Switzerland, and the United States.

> ***"The symposium's goal was to present examples of successful transition programs."***

An abstract competition was held and attracted 95 successful abstracts from around the world. The two top papers were presented by Dr. Ashwin Nathan of the Boston Adult Congenital Heart Program ("Exercise Oscillatory Ventilation in Patients with Fontan Physiology"), and by Dr. Romy Franken from the Academic Medical Center in Amsterdam ("Suppressing Inflammation in Marfan Syndrome: Be Careful!").

The social highlight of the meeting was the evening baseball game between the Cincinnati Reds and the Los Angeles

Research award presentation by Dr. Gary Webb to Dr. Romy Franken of Amsterdam.

Research award presentation by Dr. Gary Webb to Dr. Ashwin Nathan of Boston.

Strategizing by the Canadian Jeopardy team – Drs. Erwin Oechslin and Will Wilson.

Dodgers. Two hundred meeting attendees became baseball fans that night (for many of them this was their first time at a baseball game), and enjoyed a visit from the Cincinnati Reds mascots. Everyone joined

The Dutch team triumphs at International Jeopardy – Drs. Barbara Mulder and Folkert Meijboom.

voices in the singing of "Take Me out to the Ballgame" on the Jumbotron during the seventh inning stretch.

The entertainment highlight of the meeting was the International Jeopardy Competition sponsored by The International Society for Adult Congenital Heart Disease, and hosted by the always amazing Curt Daniels. Teams from the Netherlands, Switzerland, Canada and the United States competed ferociously for the grand prize, a giant trophy. The Dutch swept the competition, bringing home not just the trophy, but also pride for their country!

The meeting was recorded and will be available to everyone in a few months on the ACHD Learning Center site (www.learningcenter.org).

Next year's meeting will be held in Toronto, Canada, June 3rd-6th, 2015.

CCT

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Medical News, Products & Information

Smoking During Pregnancy May Raise Risk for Heart Defects in Babies

Women who smoke during pregnancy may be putting their newborns at risk for congenital heart defects, and the more they smoke, the higher the risk, according to a study presented, May 3rd, at the *Pediatric Academic Societies (PAS) Annual Meeting* in Vancouver, British Columbia, Canada.

Cigarette smoking during pregnancy has been linked to many birth defects, such as cleft lips and palates, and missing and deformed limbs. Some studies also suggest maternal smoking may be associated with heart defects.

The authors of this study used birth certificate data and hospital discharge records from Washington state to determine if maternal smoking during the first trimester of pregnancy is linked to heart defects and if so, what types of defects.

"I care for kids with complex congenital heart disease on a daily basis, and I see these kids and their families enduring long hospitalizations and often sustaining serious long-term complications as a result of their disease. Usually, the cause of a heart defect is unknown. I saw this research as an opportunity to study what might be a preventable cause of congenital heart defects," said lead author Patrick M. Sullivan, MD, FAAP, clinical fellow in pediatric cardiology at Seattle Children's Hospital and a master's student in epidemiology at the University of Washington School of Public Health.

Using hospital discharge records, researchers identified 14,128 children born with a variety of heart defects from 1989-2011. They matched these cases to 62,274 children without heart defects born in the same year. Then, they compared the proportion of children with heart defects whose mothers reported smoking during pregnancy to the proportion of children without heart defects whose mothers smoked. Mothers' smoking status, as well as how much they smoked daily, was available from birth certificates.

Results showed that children with heart defects were more likely than those without heart defects to have been born to mothers who smoked, and the risk was highest in the heaviest smokers. In addition, although women 35-years-of-age and older were less likely to smoke during pregnancy than younger women, older women had a higher risk of having a child with a heart defect if they smoked.

Newborns whose mothers smoked were at about a 50 to 70% greater risk for anomalies of the valve and vessels that carry blood to the lungs (pulmonary valve and pulmonary arteries) and about a 20% greater risk for holes in the wall separating the two collecting chambers of the heart (atrial septal defects). All of these defects often require invasive procedures to correct.

Researchers also found that in recent years about 10% of women giving birth reported smoking during pregnancy. They estimated that maternal smoking during the first trimester may account for 1% to 2% of all heart defects.

"Women, particularly younger women, are still smoking while pregnant, despite largely successful public health efforts to reduce smoking in the general public over the past few decades," Dr. Sullivan concluded. "Ongoing cigarette use during pregnancy is a serious problem that increases the risk of many adverse outcomes in newborns. Our research provides strong support for the hypothesis that smoking while pregnant increases the risk of specific heart defects."

Discovery That Heart Cells Replicate During Adolescence Opens New Avenue for Heart Repair

It is widely accepted that heart muscle cells in mammals stop replicating shortly after birth, limiting the ability of the heart to repair itself after injury. A study published by Cell Press May 8th in the journal *Cell* now shows that heart muscle cells in mice undergo a brief proliferative burst prior to adolescence, increasing in number by about 40% to allow the heart to meet the increased circulatory needs of the body during a period of rapid growth. The findings suggest that thyroid hormone therapy could stimulate this process and enhance the heart's ability to regenerate in patients with heart disease.

"We not only challenge 120-year-old dogma by showing that cardiac muscle cells are capable of extensive replication well into early preadolescence, but we also identify endocrine and local growth factors that could facilitate this process," says senior study author Ahsan Husain of the Emory University School of Medicine. "In the future, regenerative heart therapies in children may be possible by directly activating replication of cardiac muscle cells, without having to administer stem cells to the heart."

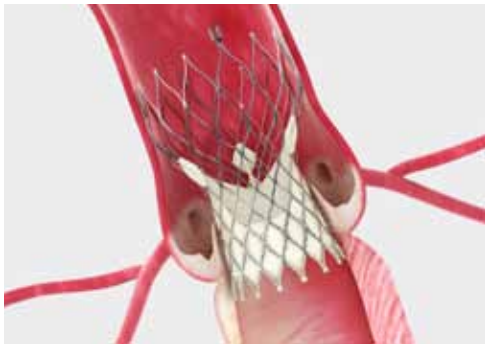
Past research has suggested that heart muscle cells stop dividing by the time mice are about one week old. But the heart grows substantially in mammals between birth and adolescence to accommodate an increase in the growing body's need for oxygenated, nutrient-rich blood. Heart muscle cells grow in size during preadolescence, but only to a limited degree, so the cellular processes accounting for the heart's rapid growth during this period have been unclear.

Shedding light on this mystery, a research team led by Husain, Nawazish Naqvi of the Emory University School of Medicine, and Robert Graham of the Victor Chang Cardiac Research Institute found that preexisting heart muscle cells retain their ability to replicate until well after birth. These cells undergo an intense 24-hour spurt of cell division in preadolescent 15-day-old mice, increasing in number by about half a million or 40%. This process was initiated by a surge in blood levels of thyroid hormone, which activated a signaling pathway known to trigger cell proliferation. Moreover, the ability of the heart to recover after injury was enhanced in the period before adolescence, corresponding to approximately 8 to 10 years of age in humans, compared with later in development.

"The brevity of this proliferative burst could explain why it has previously gone undetected," Graham says. "We may be able to take advantage of this window of opportunity, for example, by giving thyroid hormone to improve repair of the heart in babies born with heart defects, or to reactivate heart muscle cells in adults who experience a heart attack later in life."

Accelerated Approval Obtained after Clinical Outcomes with Self-Expanding Valve Prove Superior to Surgical Aortic Valve Replacement at One Year - CoreValve System Now Available to More U.S. Patients than any other Transcatheter Aortic Valve

In mid-June Medtronic, Inc. (www.medtronic.com) announced the U.S. Food and Drug Administration (FDA) approval



Medtronic CoreValve® System - sits within the same area as the native valve. Image Courtesy of Medtronic, Inc.

of the self-expanding transcatheter CoreValve® System for patients with severe aortic stenosis who are at high risk for surgery. This approval was based on groundbreaking research that showed clinical outcomes at one year with the CoreValve System were superior to open-heart surgery, the current gold standard for aortic valve replacement.

The FDA approved the CoreValve System without the need for an independent device advisory panel review due to its exceptionally positive clinical results demonstrated in the High Risk Study of the CoreValve U.S. Pivotal Trial. The head-to-head study, comparing transcatheter aortic valve replacement (TAVR) with the CoreValve System to traditional surgical aortic valve replacement, met its primary endpoint with high survival at one year for patients receiving the CoreValve System (85.8%), which was statistically superior to patients receiving a surgical valve (80.9 %).

"This rigorous trial has defined a new standard for transcatheter valve performance, with superior results that give physicians even more confidence in making TAVR treatment decisions," said David H. Adams, MD, Chair of the Department of Cardiothoracic Surgery at the Mount Sinai Hospital, New York City, national co-principal investigator of the CoreValve U.S. Pivotal Trial. "With this approval, we can treat more patients due to the broad range of CoreValve sizes, and we have an option compared to surgery that provides a greater chance for a longer life while minimizing the risk of stroke."

For patients treated with the CoreValve System in the High Risk Study, rates of stroke - one of the most concerning complications of valve replacement because it increases the risk of death and can have a dramatic impact on quality of life - were low and not statistically different than rates experienced by surgery patients. The rate of MACCE (major adverse cardiovascular or cerebral events) was significantly better for CoreValve patients at one year, and overall hemodynamic (blood flow) performance was better in CoreValve patients than in surgical patients across all time points.

The CoreValve System was designed to serve the clinical needs of the broadest range of patients with aortic stenosis. The valve's self-expanding frame provides controlled deployment, enabling physicians to accurately place the valve inside a patient's original valve, while conforming to the native annulus to provide a seal. The FDA approved the entire CoreValve platform - including the 23mm, 26mm, 29mm and 31mm size valves - all of which are delivered through the smallest commercially available TAVR delivery system (18Fr, or approximately 1/4 inch), making it possible to treat patients with difficult or small vasculature.

"It's rewarding that we can now offer this life-saving therapy to patients at increased risk for surgery," said John Liddicoat, MD, Senior VP, Medtronic, and President of the Medtronic Structural Heart Business. "There is a lot of excitement among U.S. heart teams for the CoreValve System's high risk approval, and its unique design that leads to the clinical outcomes seen in the High Risk Trial. We will continue to safely introduce CoreValve System to these physicians, supporting heart teams through comprehensive training and education, imaging and patient evaluation programs."

Medtronic worked closely with the FDA throughout the pivotal clinical trial and Pre-Market Approval (PMA) review process. FDA granted Priority Review Designation for both the Extreme Risk and High Risk PMA submissions. Priority designation is granted to new therapies of major public health interest. Based on the strength of the trial data, FDA determined that sufficient information was available to evaluate the safety and efficacy of the Medtronic CoreValve System for both patient groups without the need for external Advisory Committee panels. This milestone, along with the priority review designation, accelerated regulatory approvals for the life-saving device.

The CoreValve System was approved by the FDA for patients at extreme risk in January 2014. Since receiving CE (Conformité Européenne) Mark in 2007, the CoreValve System has been implanted in more than 60,000 patients in more than 60 countries.

For more information about the CoreValve System, call 877-526-7890 or go to www.corevalve.com.

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