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

November 2016; Volume 14; Issue 11
International Edition

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Case Report: Multiple Genetic Mutations in an Infant with Refractory Brugada-Like Syndrome

By Katie Kowalek, MD; Ricardo A. Samson, MD; Kevin Engelhardt MD; Yung Lau, MD; Santiago O. Valdes, MD

Abbreviations: Brugada Syndrome (BrS); Right Bundle Branch Block (RBBB); beats per minute (bpm); electrocardiogram (ECG); ventricular tachycardia (VT); atrioventricular (AV); cardiopulmonary resuscitation (CPR); extracorporeal membrane oxygenation (ECMO), implantable cardioverter defibrillator (ICD).

Introduction

Brugada Syndrome (BrS) was first described by Pedro and Josep Brugada in 1992 as Right Bundle Branch Block (RBBB) with ST segment elevation, conveying increased risk of sudden cardiac death.^{1,2} Over time, BrS has been found to be associated with gene mutations encoding for cardiac sodium, calcium, or potassium ion channels.³ These channelopathies predominate mostly in the epicardium, resulting in a loss of function of sodium or calcium channels, or a gain of function in potassium channels.

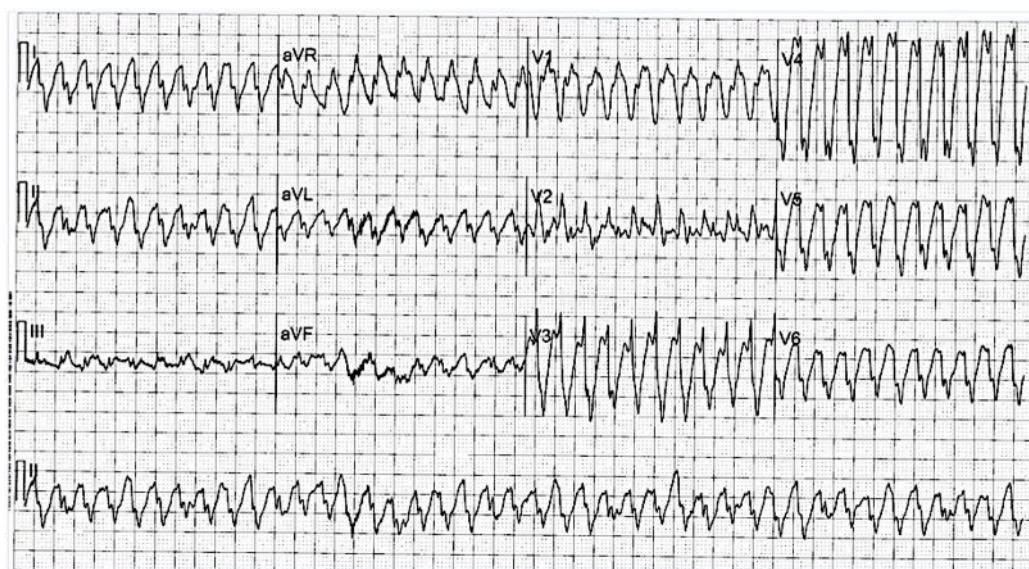


Figure 1. Initial 12-lead ECG showing ventricular tachycardia 252 bpm with RBBB morphology.

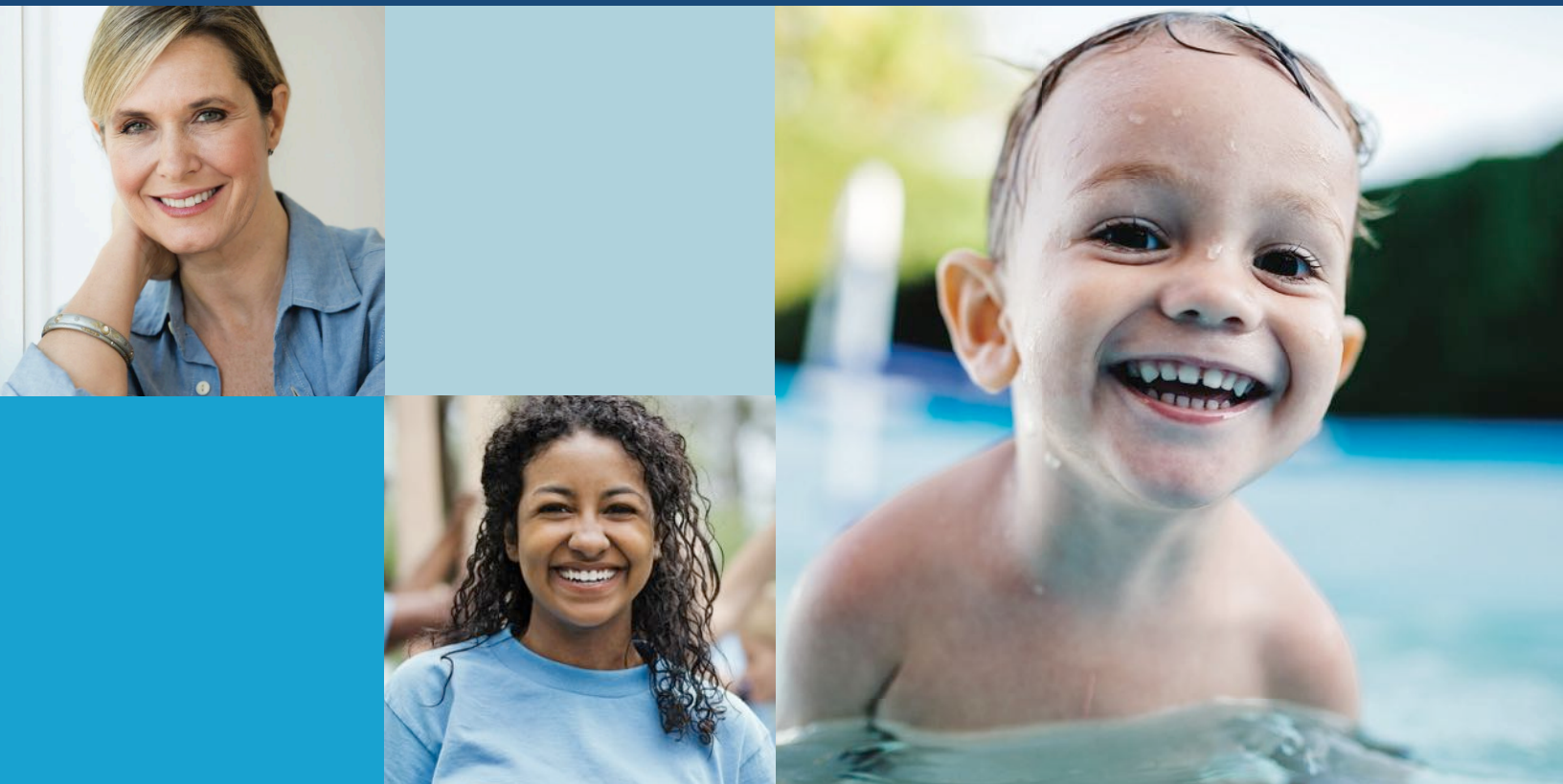
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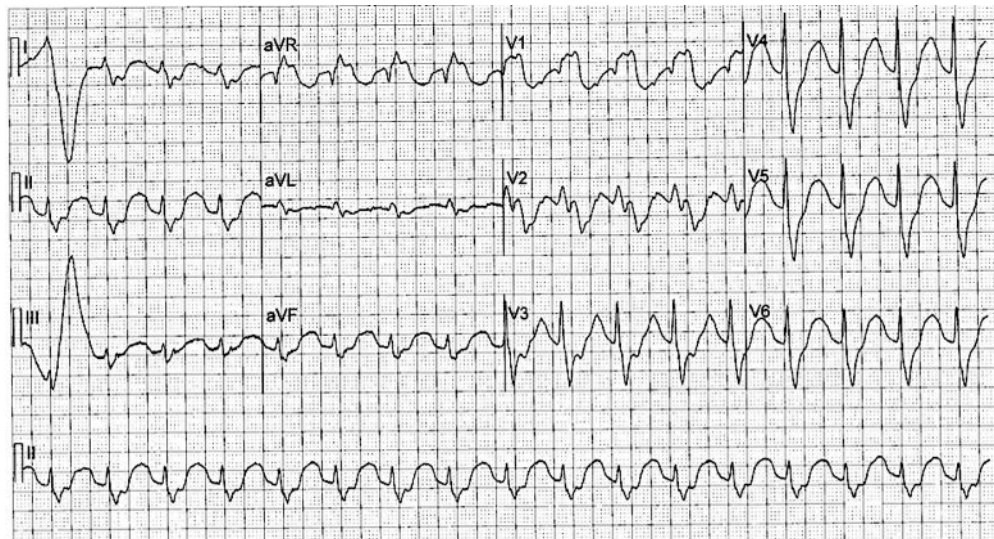


Figure 2. After receiving two boluses of IV amiodarone, 12-lead ECG shows sinus rhythm 102 bpm, with PR interval 176 ms, and QRS duration of 172, RBBB morphology with ST segment elevation in lead V1.

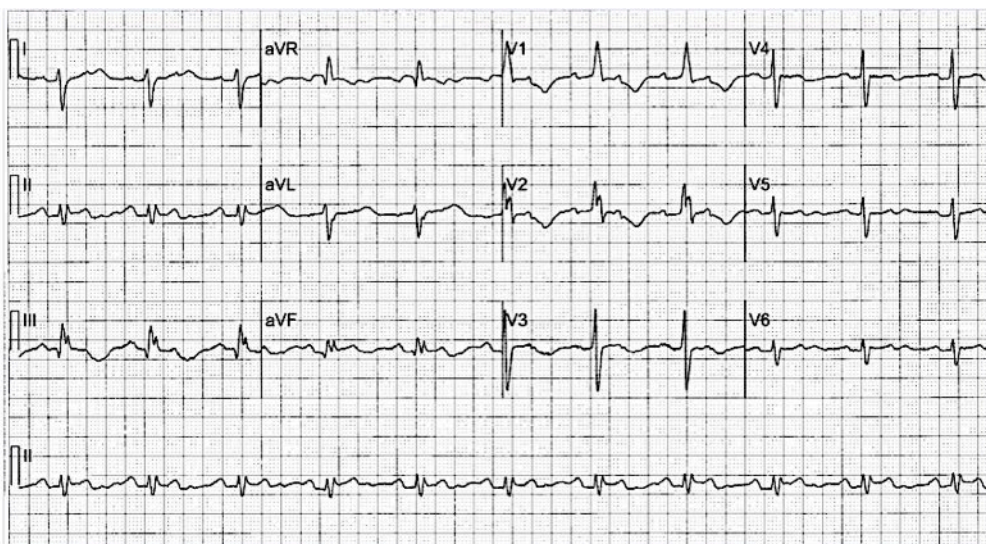


Figure 3. On hospital Day 2, 12-lead ECG shows sinus rhythm with 2:1 AV block, RBBB, and prolonged PR interval and QTc. He had been on an amiodarone drip for approximately 12 hours at this point.

Consequently, during early depolarization (Phase I of the cardiac action potential), decreases in I_{Na} or increases in I_{to} cause a transmural voltage gradient which leads to the characteristic electrocardiographic changes of BrS.¹ While BrS is more commonly diagnosed in adults, it is rare in children.⁴ Additionally, the typical BrS ECG pattern may not be observed in infants. As described in a case series by

Kanter et al⁴, 4 out of 5 infants with genetic mutations encoding for BrS did not demonstrate classic RBBB with ST elevation. They used the term Brugada-like Syndrome to describe this group of patients. We present an infant with refractory ventricular tachycardia who was found to have three distinct mutations encoding for cardiac ion channels consistent with Brugada-like Syndrome.

Case Report

A 39-day-old male presented to the hospital for fever and dyspnea associated with perioral cyanosis. Admission examination was significant for tachypnea with upper airway congestion and auscultation revealing coarse breath sounds. Cardiac examination revealed a normal S1/S2 without murmurs, rubs, or gallops, and strong distal pulses. Abdominal exam was benign without hepatomegaly.

Shortly after receiving an albuterol treatment for wheezing and hypoxia, he developed tachycardia with a rate of up to 300 beats per minute (bpm). He was hemodynamically stable. An electrocardiogram (ECG) revealed a wide-complex tachycardia, rate 252 bpm with RBBB morphology and QRS axis of -126° (Figure 1). Administration of adenosine did not convert his tachycardia, but did reveal atrioventricular (AV) dissociation. He was given two IV boluses of amiodarone, and placed on a continuous infusion. Within 6 hours, he converted to sinus rhythm, 102 bpm with wide QRS (172 ms) and RBBB morphology in lead V1 (Figure 2). Thereafter, however, sustained Ventricular Tachycardia (VT) recurred and persisted, during which time only rate control could be achieved. He received multiple boluses of IV amiodarone, and the continuous infusion was increased to 15 mcg/kg/min. Eventually, the VT terminated, with resulting sinus rhythm with first degree AV block and narrow QRS alternating with wide QRS in a 2:1 to 3:1 fashion. On hospital Day 2, he developed 2:1 AV block with associated bradycardia (Figure 3), hypotension, and poor perfusion. Vasopressin was started, amiodarone was weaned, and he was placed on IV lidocaine infusion up to 40 mcg/kg/min. Seizures developed secondary to lidocaine toxicity and breakthrough VT occurred as lidocaine was decreased; therefore, an esmolol infusion was added. By hospital Day 3, he continued to have intermittent non-sustained VT with resultant hypotension and worsening metabolic acidosis. When his VT converted, he was significantly bradycardic; therefore, a temporary transvenous pacing catheter was placed. On hospital Day 7, he once again developed sustained VT with hypotension and poor perfusion. Despite resuscitative efforts, he became bradycardic and pulseless; cardiopulmonary resuscitation (CPR) was initiated and advanced life support given. After one hour of chest compressions, he was emergently placed on venous-arterial Extracorporeal Membrane Oxygenation



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(ECMO). He remained on ECMO for four days, during which time he returned to sinus rhythm with frequent ventricular ectopy on lidocaine, esmolol, and vasopressin infusions. Because of continued significant bradycardia associated with his anti-arrhythmic therapy, permanent dual chamber pacemaker placement was performed. Initially, an attempt at surgical placement of epicardial pacemaker was unsuccessful because sites with satisfactory pacing thresholds could not be found. A second attempt with wider exposure of the epicardium was successful. Eventually he was transitioned to oral propranolol, amiodarone, and mexiletine. While he continued to have short runs of VT, he did not have any hemodynamic instability and was discharged home.

DNA for genetic testing of cardiac ion channel mutations was sent in the first week of hospitalization and results returned after discharge. He was found to have two distinct Class I mutations – SCN5A Arg225Trp, SCN5A Val845fs – and a third Class II mutation, SCN5A Thr630Met.


At 5 months of age, this patient and his family relocated. Based on the results of the DNA testing, amiodarone was discontinued, and quinidine was started. He continued on mexiletine and propranolol. Unfortunately, he continued to have incessant non-sustained VT. An Implantable Cardioverter Defibrillator (ICD) with epicardial leads was placed, during which he was noted again to have high pacing thresholds. He had multiple admissions for un-responsiveness with no documented arrhythmias, and had developed seizure activity that was confirmed by electroencephalogram. Levetiracetam was started to control seizure activity.

He had multiple echocardiograms throughout his course. Those in the first 5 months of life showed normal cardiac anatomy with a Patent Foramen Ovale (PFO). Beyond 5 months, echocardiograms showed a dilated left ventricle with left-ventricular diastolic dimensions greater than the 99 percentile for body surface area, which never recovered on subsequent echocardiograms. Ejection fractions ranged from 30% to 70%.

At 10 months of age, the patient developed a seizure associated with profound bradycardia. He had subsequent loss of pacer capture and required CPR and advanced life support. He was placed on ECMO, from which he could not be weaned, and he unfortunately died.

Discussion

Brugada Syndrome has been previously described in infants and children in scattered case reports and one case series.⁴ The incidence of BrS is difficult to determine, but is estimated to be 1-5 per 10,000.⁵ In adults, BrS is more common in men; however, in children, there seems to be no sex predilection.⁶ Whereas, the classic ECG findings of RBBB with ST segment elevation in lead V1 is often noted in adults with BrS, these ECG features may not be present in infants. Kanter et al,⁴ described 5 infants with genetic-proven BrS-causing mutations, but only one patient manifested the typical RBBB and ST elevation. They reasoned that normal developmental changes in cardiac ion channel density and function could contribute to the lack of a classic BrS ECG pattern. They also surmised that the relatively higher balance of right-ventricular to left-ventricular mass in infants compared to mature patients might



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also have influence on the surface ECG. As such, they proposed the term Brugada-like Syndrome to apply to these patients who may have a "heterogeneous electrophysiologic milieu."

The clinical presentation of infants with BrS is variable. Skinner et al reported a 21 month-old child with recurrent febrile seizures, who was found to have VT during these episodes consistent with BrS. Her seizures were felt to be a result of poor cerebral perfusion during episodes of VT.⁷ Other case reports describe aborted sudden cardiac death,^{7,8} recurrent episodes of cyanosis with crying,⁹ and febrile upper respiratory infection with signs of low cardiac output on exam.¹⁰ A common presentation, such as in our patient, is VT that occurs with fever.⁶

Our patient also had the characteristic of high pacing thresholds noted on two surgical procedures for pacemaker and ICD implantation. Such a finding was previously reported by Lopez et al in a patient with homozygous mutation for SCN5A and in Kanter's series, 3 infants had high ventricular pacing thresholds.^{11,4} Of interest, Lopez demonstrated poor atrial capture thresholds, Kanter demonstrated poor ventricular thresholds, and our patient had both poor atrial and ventricular capture thresholds. To our knowledge, no other reports have described poor atrial and ventricular capture thresholds in patients with SCN5A mutations.

A review of the literature reveals that multiple mutations in the SCN5A gene in the same person are very uncommon. Barajas-Martinez et al reported a 45 year-old man shown to have lidocaine-induced BrS ECG findings. Genetic analysis revealed a double-mutation in SCN5A (Val232Ile and Leu1308Phe).¹² Nof et al described a 16 year-old male with bradycardia and RBBB, found to have three mutations in SCN5A (Val1251Met, Val1924Thr, and Lys1492del).¹³ An international multi-center retrospective analysis by Kapplinger et al revealed a small cohort of patients with two SCN5A mutations. It was noted that these patients were younger at diagnosis than the remainder of the cohort (29.7+/-16.2 vs 39.2+/-14.4). However, the youngest patient in that cohort was 2 years of age.¹⁴ There are no reports of multiple genetic mutations in SCN5A in infants to our knowledge.

Our patient had genetic analysis completed by Transgenomic, Inc. (New Haven, CT). The results are categorized by class. Class I mutations are expected to predispose to disease. Class II mutations may predispose to genetic heart disease. However, it has not been fully shown to be of significance. Neither Class I nor II mutations have been found in healthy controls in the general population. Class III mutations have been found in healthy controls and are not thought to be disease-causing mutations. Our patient's testing revealed two Class I mutations (SCN5A: Arg225Trp, Val845fs), one Class II mutation (SCN5A: Thr630Met), and three Class III mutations (CACNA1C: Leu1868Pro; SCN1B: Leu210Pro; SCN5A: His558Arg). The SCN5A Arg225Trp mutation is a point mutation, resulting in an amino acid change within the transmembrane region of the SCN5A protein. In vitro analysis of this missense mutation caused a 90% reduction in INa in sodium channels expressed in *Xenopus* oocytes. While this mutation has been reported in patients suspected of having Long QT Syndrome and Cardiac Conduction Disease, Kapplinger et al have also observed this mutation in three unrelated probands suspected of having BrS.¹⁴ The SCN5A Thr630Met has not been reported in the



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literature, but has not been observed in healthy controls. The Val845fs mutation is a frameshift mutation which likely results in abnormal termination of the SCN5A protein in Domain II, transmembrane region 5, and would be expected to have deleterious effects on sodium channel function. It too has been reported in one patient suspected of having BrS.^{14,15}

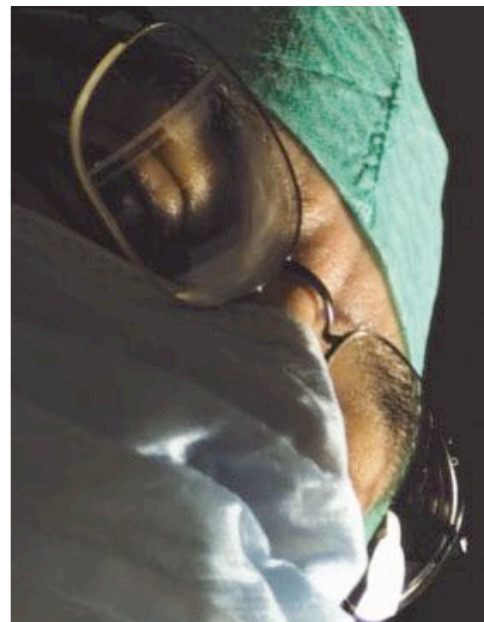
We surmise that the likelihood of our patient having simultaneous spontaneous mutations to account for his genotype is exceedingly low and, thus, it is much more likely that he inherited one or more of these mutations from his mother and/or father. Thus, we speculate that he would have received one Class I mutation from his mother and the other Class I mutation from his father, and that the combination of the two mutations would have rendered his cardiac sodium channels so dysfunctional so as to significantly decrease INa and convey a poor prognosis. However, given the complexity of his social situation, obtaining biological samples from family members was not able to be performed.

Conclusions

In summary, we present an interesting case of an infant with Brugada-like Syndrome who presented with fever, refractory ventricular tachycardia, and high pacing thresholds. He did not have the typical ECG pattern of BrS, but was ultimately found to have three mutations significant for BrS. To our knowledge, this has yet to be described in an infant. Unlike many previous reports of infants with BrS, our patient had an extremely difficult course with refractory ventricular arrhythmias. The acuity of his course and ultimate demise may have been related to the multiple mutations in the SCN5A gene, resulting in marked aberrations in the function of this ion channel.

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Medical News, Products & Information

Compiled and Reviewed by Tony Carlson,
Senior Editor

Edwards INSPIRIS RESILIA Valve Receives CE Mark

PRNewswire -- Edwards Lifesciences Corporation, announced in late September, it received CE Mark for its INSPIRIS RESILIA Aortic Valve, the first in a new class of resilient heart valves. Incorporating the advanced RESILIA tissue, the INSPIRIS valve leverages features of the trusted PERIMOUNT Magna Ease Valve and includes the proprietary VFit technology, which is designed for potential future valve-in-valve procedures.



INSPIRIS valve with inside view of VFit technology.

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"Heart valve patients are living longer, and want to maintain their active lifestyles. The INSPIRIS valve provides surgeons and their patients with an advanced treatment option that represents an ideal foundation for the future treatment of heart valve patients," said Vinayak (Vinnie) Bapat, MBBS, MS, FRCS, FRCS.CTh, consultant cardiothoracic surgeon at Guy's and St. Thomas' Hospital in London.

Bernard Zovighian, Edwards' Corporate VP, Surgical Heart Valve Therapy, said, "The INSPIRIS valve creates a new class of surgical valves and includes key patient-focused innovations, such as the resilient tissue and the VFit technology, to advance the treatment of patients with heart valve disease. In partnership with surgeons, Edwards is

committed to developing differentiating technologies that set a new standard for surgical heart valves."

A key innovation of the INSPIRIS valve is RESILIA tissue, a new, first-of-its-kind tissue platform that has been in development for more than a decade and builds on Edwards' 40 years of leadership in tissue technology. RESILIA tissue utilizes Edwards' integrity preservation technology, which preserves the tissue and provides improved anti-calcification properties and sustained hemodynamic performance. In addition, the valve is stored dry and ready to use.

CE Mark of the INSPIRIS valve was supported by the COMMENCE pivotal trial, a global, U.S. Food and Drug Administration premarket approval study. One-year results of this study were presented during this year's late-breaking sessions at the American Association for Thoracic Surgery's (AATS) 96th annual meeting. The results of 673 patients showed no cases of structural valve deterioration, valve thrombosis or nonstructural valve dysfunction. In addition, a series of pre-clinical studies have shown RESILIA tissue to offer key benefits, such as significantly reduced calcification and sustained hemodynamics compared to a current treatment option.

Additional data from the COMMENCE trial was presented Oct. 3rd during a late-breaking session at the 30th European Association for Cardio-Thoracic Surgery's (EACTS) annual meeting. In addition, three-year results of the RESILIA tissue EU feasibility study was presented during EACTS on Oct. 2nd.

The INSPIRIS valve is planned for a targeted commercial release in Europe in Q4 of 2016, and a full launch in Q1 of 2017. The VFit feature is available in sizes 19mm through 25mm. *It is not approved for commercial use in the U.S.*

Edwards Lifesciences is based in Irvine, California. For more information, visit www.edwards.com.

Novel Heart Valve Replacement Offers Hope for Thousands with Rheumatic Heart Disease

A novel heart valve replacement method was revealed that offers hope for the thousands of patients with Rheumatic Heart Disease

(RHD) who need the procedure each year. The research was presented at the SA Heart Congress 2016 on September 6th, 2016.

The Annual Congress of the South African Heart Association was held in Cape Town from 8th to 11th September 2016, and was jointly organised with the annual congress of the World Society of Cardiothoracic Surgeons. Experts from the European Society of Cardiology (ESC) presented a special programme.

"Over the past decade, heart valve surgery has been revolutionised by Transcatheter Aortic Valve Implantation (TAVI)," said lead author Dr. Jacques Scherman, a cardiac surgeon in the Chris Barnard Division of Cardiothoracic Surgery, University of Cape Town, South Africa. "Heart valves are replaced or repaired via a catheter, obviating the need for open heart surgery or a heart-lung machine."

He continued, "TAVI is only indicated in patients with Calcific Degenerative Aortic Valve Disease, which is the most prevalent aortic valve pathology in developed countries. In developing countries, Rheumatic Heart Disease still accounts for the majority of patients in need of a heart valve intervention."

Rheumatic Heart Disease is caused by rheumatic fever, which results from a streptococcal infection. Patients develop fibrosis of the heart valves, leading to Valvular Heart Disease, heart failure and death. In Africa alone there are around 15 million patients living with Rheumatic Heart Disease of whom 100,000 per year might need a heart valve intervention at some stage of their life. The vast majority of these patients have no access to cardiac surgery or sophisticated cardiac imaging.

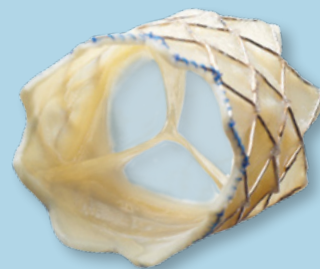
Dr. Scherman said, "Inspired by the success of TAVI for Calcific Aortic Valve Disease, we developed a simplified TAVI device for transcatheter aortic valve replacement in patients with RHD."

Currently available, balloon expandable TAVI devices require the use of sophisticated cardiovascular imaging to correctly position the new valve. They also use a temporary pacemaker which allows the heart to beat so quickly that it stops blood circulating to the rest of the body (called rapid ventricular pacing).



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The Melody TPV System received Health Canada approval in December 2006
and US approval under an HDE on January 25, 2010 (H080002).
PMA approval received January 27, 2015 (P140017).

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Melody™ Transcatheter Pulmonary Valve Ensemble™ II Transcatheter Valve Delivery System

Important Labeling Information for United States

Indications: 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
 - regurgitation: \geq moderate regurgitation, AND/OR
 - stenosis: mean RVOT gradient \geq 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 suggests that 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. Standard medical and surgical care should be strongly considered in these circumstances.
- 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, blistering, or peeling of skin, pain, swelling, or bruising at the catheterization site.

Potential device-related adverse events that may occur following device implantation include the following: stent fracture, *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.

*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

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.

Important Labeling Information for Geographies Outside of the United States

Indications: The Melody Transcatheter Pulmonary Valve is indicated for use in patients with the following clinical conditions:

- Patients with regurgitant prosthetic Right Ventricular Outflow Tract (RVOT) conduits with a clinical indication for invasive or surgical intervention, OR
- Patients with stenotic prosthetic RVOT conduits where the risk of worsening regurgitation is a relative contraindication to balloon dilation or stenting.
- Existence of a full (circumferential) RVOT conduit that was equal to or greater than 16 mm in diameter when originally implanted.

The intended lifetime for the Melody device is 2 years.

Contraindications:

- Venous anatomy unable to accommodate a 22 Fr size introducer sheath; implantation in left heart.
- Unfavorable right ventricular outflow tract for good stent anchorage.
- Severe right ventricular outflow obstruction, which cannot be dilated by balloon.
- Obstruction of the central veins.
- Clinical or biological signs of infection.
- Active endocarditis.
- Known allergy to aspirin or heparin.
- Pregnancy.

Potential Complications/Adverse Events: 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.

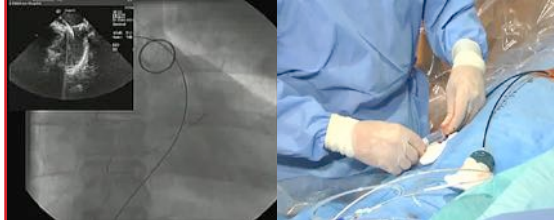
Potential device-related adverse events that may occur following device implantation include the following: stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis,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

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

The Melody Transcatheter Pulmonary Valve and Ensemble II Transcatheter Delivery System has received CE Mark approval and is available for distribution in Europe.

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- Intended Intervention - ASD Closure - *Live Case*
- and many more....

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**CONGENITAL CARDIOLOGY
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Dr. Scherman said, "Rapid ventricular pacing can only be tolerated for a short period and, therefore, limits the time available to do the implantation."

The team in South Africa developed a novel TAVI device which is "non-occlusive", meaning that there is no need to stop blood circulating to the body with rapid ventricular pacing. The device is also "self-locating," and does not require sophisticated cardiac imaging for positioning.

The proof-of-concept study presented tested the device in a sheep model. The investigators found that the device was easy to use and positioned the valve correctly, and the procedure could be performed without rapid ventricular pacing.

Dr Scherman said, "We showed that this new non-occlusive, self-locating TAVI delivery system made it easy to perform transcatheter aortic valve replacement. Using tactile feedback the device is stabilised in the correct position within the aortic root during the implantation. It also has a temporary backflow valve to prevent blood leaking backwards into the ventricle during the implantation of the new valve. All these factors together allowed for a slow, controlled implantation compared to the currently available balloon expandable devices."

He added, "This simplified approach to transcatheter aortic valve replacement could be done in hospitals without cardiac surgery at a fraction of the cost of conventional TAVI. It has the potential to save the lives of the large numbers of Rheumatic Heart Disease patients in need of valve replacement."

Professor Karen Sliwa, President of the South African Heart Association, said, "I am truly excited that we have not only an internationally strong group working on epidemiology and prevention of RHD at the University of Cape Town, but also a dedicated and successful surgical group, led by Prof. Peter Zilla at the Chris Barnard Department. Although prevention is the final goal, millions will need surgery as a life-saving measure for decades to come. Knowing from my own Pan-African collaborations how inadequate the provision of cardiac surgery is on the African continent this fascinating solution promises surgical help for all these young patients with Rheumatic Heart Disease on a continent that has a fair density of general hospitals but hardly offers any open heart surgery."

Professor Fausto Pinto, ESC President & Course Director of the ESC Programme in South Africa, said, "The development of innovative therapeutic strategies is extremely important to allow a larger number of patients to be treated."

Is It OK for a Doctor to Attend a Patient's Funeral?

Newswise — New research at the University of Adelaide has shed light on how many doctors are attending the funerals of their patients, and the reasons behind their choice. The researchers say more needs to be done within the medical profession to openly discuss the issue.

In a study published online ahead of print in the journal *Death Studies*, researchers from the University's School of Psychology and School of

Medicine report on the practices and attitudes towards funeral attendance of more than 430 Australian doctors. The publication is part of a nationwide survey of more than 1,000 health professionals.

"Our survey was aimed at better understanding what motivates health professionals to attend their patients' funerals, what barriers they may experience in attending, and their attitudes towards the issue of funeral attendance," says Dr. Sofia Zambrano, who conducted this work as a follow-up to her PhD in the School of Medicine at the University of Adelaide.

The survey found that 57% of the doctors surveyed had attended at least one funeral of a patient – but the number varied greatly depending on which medical specialisation they had pursued. For example, 71% of general practitioners had attended a patient's funeral; 67% of oncologists; 67% of psychiatrists; 63% of palliative medicine specialists; 52% of surgeons and 22% of intensive care specialists.

"The death of a patient can be a very emotional and isolating experience for physicians, and some may regard it as the ultimate failure of their professional care," says Associate Professor Greg Crawford, study co-author and Associate Professor of Palliative Medicine in the University's School of Medicine.

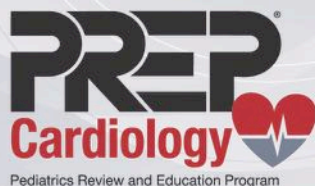
He says the benefits of attendance may be twofold, "Funeral attendance seems to be a practice that may help physicians deal with their emotions after a patient dies, and in turn, it can also be of comfort for the patient's family.

"However, there are differing views within medicine about whether or not it's acceptable to attend a patient's funeral, with some doctors seeing it as 'unprofessional', and others feeling that their colleagues would disapprove of them attending, which in fact were factors associated to non-attendance to funerals in our study," Associate Professor Crawford says.

The study also found that female doctors were more likely to attend a patient's funeral than their male counterparts, were more open to crying and expressing grief at the funeral, and they actively discussed attending patients' funerals with their colleagues and families. Those who were least likely to attend were young male doctors with fewer years of medical experience.

Dr. Zambrano says that because the decision is a personal one, the paper's authors have refrained from advocating attendance or non-attendance at funerals. "We aim to contribute to a more open discussion about this poorly researched topic, and to provide a clearer picture of actual practices and attitudes of a large sample of physicians and other health professionals," she says.

"The role of peer perception, and the hesitation of doctors to discuss funeral attendance and death more broadly with colleagues are important issues to consider. The medical community should ask itself whether funeral attendance needs to – and can – be addressed more openly, whether death and dying should be discussed more candidly among health professionals, and what effects these discussions may have on job satisfaction and on the mental health of medical practitioners."



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Childhood Hypertension Associated with Cognitive Issues

Hypertension, more commonly known as high blood pressure, has increased significantly in children, paralleling the current childhood obesity epidemic. Although we know that adult hypertension can affect the brain, little research has been done on the cognitive effects of childhood hypertension. In a new study scheduled for publication in *The Journal of Pediatrics*, researchers found that hypertension is associated with cognitive issues in children and adolescents.

Marc B. Lande, MD, MPH, and researchers from the University of Rochester, Emory University, Maimonides Medical Center, University of Texas at Houston, University of North Carolina, Thomas Jefferson University, University of Maryland, and the University of California at Los Angeles, compared different tests of cognitive skills between 75 10-18-year-old children with newly-diagnosed hypertension, and 75 matched children without hypertension. Children who had other factors that are known to affect cognitive skills were excluded from the study (e.g., ADHD, learning disabilities, sleep disorders). According to Dr. Lande, "We wanted to make sure that if we found differences between children with and without hypertension, it was likely associated with the hypertension itself, not any of these other factors."

The researchers found that the children with hypertension performed worse on the cognitive tests that measured visual and verbal memory, processing speed, and verbal skills. Additionally, more children with sleep issues had hypertension, which intensified the effect of poor sleep on cognition and executive function. It is important to note that the differences between groups were small and that the average cognitive test scores of both groups were largely within normal ranges. The children with hypertension were not cognitively impaired, but rather performing less well than children without hypertension.

Overall, this study provides evidence that hypertension in children is associated with a subtle pattern of decreased performance on cognitive testing. Notes Dr. Lande, "In the future, we want to better understand if there

are physical changes to the brain in children who have hypertension that could explain these cognitive test results." Knowing how these physical changes might affect cognitive skills could be important in future studies that assess whether antihypertensive treatments could improve cognitive performance in children with hypertension and reverse or prevent future adult hypertension-related problems.

Upcoming Medical Meetings

AIMed

Dec. 12-15, 2016; Laguna Niguel, CA USA
Aimed-mi3.com

Pediatric and Adult Interventional Cardiac Symposium (PICS 2017)

Jan. 16-19, 2017; Miami Beach, FL USA
www.picsymposium.com

Sports Cardiology & Sudden Cardiac Arrest in the Young

Jan. 20-21, 2017; Anaheim, CA USA
www.choc.org/events/sudden-cardiac-arrest-young-2016/

PEDIRHYTHM VII: Pediatric and Congenital Rhythm Congress

Feb. 4-7, 2017; Thessaloniki, Greece
www.pedirhythm.org/

Cardiology 2017

Feb. 22-26, 2017; Orlando, FL
www.chop.edu/events/
cardiology-2017#.V-WXtaO-L5U

CONGENITAL CARDIOLOGY TODAY

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Sincerely,

Gary Webb, MD
CHiP Network
215-313-8058
garywebb6@gmail.com



The CHiP Network, the Congenital Heart Professionals Network, is designed to provide a single global list of all CHD-interested professionals.



Join Us for Two Weeks to Save Children with Congenital Heart Defects!

We are looking for medical volunteers who can join us in our missions to underdeveloped countries to perform pediatric cardiac surgery and care while also training the local medical staff.

For more information and our 2016 mission schedule, please visit Babyheart.org



Skamania 2016 ACHD Innovation Past and Present

Tabitha Moe, MD

The 26th Annual International Symposium on Adult Congenital Heart Disease was held at Skamania Lodge, June 2nd-4th, 2016. As always, the meeting was an immense success, hosted by the Oregon Health Knight Cardiovascular Institute, and University of Washington ACHD Programs. It was an opportunity to rest, recharge, and get out into the great outdoors. The Symposium opened with an introduction and a future vision by Ariane Marelli, with visionary thoughts about the next era in Congenital Heart Disease (CHD) care. Glenn Tringali, ACHA National Executive Director, introduced the ongoing efforts towards Accreditation of Adult Congenital Care Centers. The year 2016 is the first year in which centers will begin to be accredited through this process. The ACHA National Meeting will be held October 7th and 8th in Orlando, FL.

A local patient, Kelly Aicher, Tetralogy of Fontan (TOF) survivor and cycling enthusiast, presented his compelling story emphasizing the need for ongoing exercise in our adult Tetralogy population. He continues to be involved in biking, and is a model for maintaining cardiovascular fitness, as well as efforts for community involvement in CHD funding. We only wish that all of our patients would take a tip from his lifestyle choices.

The Congenital Heart Futures Act is a partnership with the AHA with joint research projects. This project was originally approved with the Affordable Care Act, and continues to fund ongoing cross-disciplinary research. More research is needed as CHD's seem to be the best kept secret in the medical world – with the number of patients affected growing every year

In the spirit of improving physical fitness, Tina Kaufman, PA, presented the Fitbit Frenzy – Tips for the Provider on Patient Self-Quantification, and encouraged us to help patients understand their exercise, and encourage an ongoing increase in exercise to improve cardiovascular health. If you have not already begun to evaluate patients with their personal fitness tracking devices, not only should you familiarize yourself with the devices, but also the benefits of proactive exercise and exercise prescriptions in a paradigm shift away from the exercise limitations of the past.

Jolien Roos Hesselink presented the ROPAC Registry, the Registry on Pregnancy and Cardiac Disease. It is a multi-national registry to evaluate pregnancies in Congenital Heart Disease. The ESC constructed the database, but does not fund the database.

The ESC does the analysis of the database for ROPAC, and in order to do an analysis, you must proceed through executive committee.

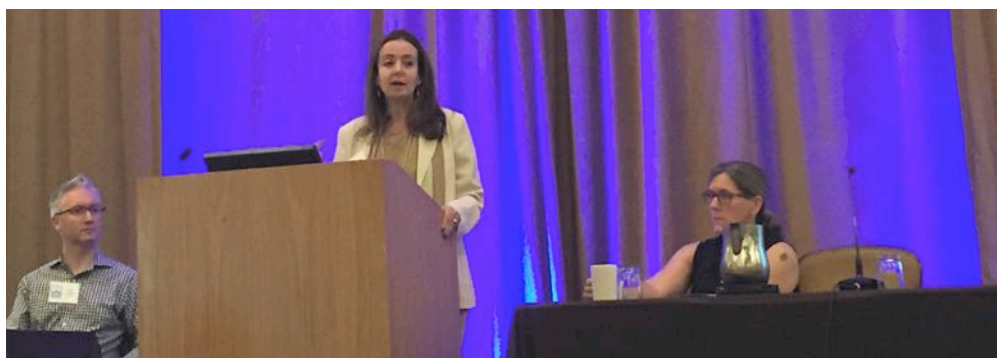
Barbara Mulder presented her experiences and inspiration to create funding for International Registries. She specifically discussed the NOTE Registry as an International, multi-centered, open prospective registry. The funding is a combination of: Professional Organizations, Private Industry and Foundations like the NIH and Public Funding through crowd sourcing. Caution with International studies, as there are time-consuming Ethical Committees in all centers! Having an adept cultural broker to navigate cultural differences and language is paramount.

Bill Davidson gave some excellent data regarding the implementation of strain imaging in the systemic RV. In contrast with the LV, the RV has less longitudinal strain and more circumferential strain, and demonstrates a wider range of values, particularly in volume overload states. There is significant vendor variability, as well as intraobserver variability in strain imaging. There may be improved RV strain values from ACEi in CCTGA, and the single RV had improved RV strain values on B-blockers.

Stephen Seslar presented CRT for CHF in TGA. The PACES/HRS Expert consensus

statement states that if dyssynchrony is not manifest in the appearance of a wide QRS, the indications for pacing are unclear. Half of single-right ventricles upgraded to Bi-V are responders. It may be possible to optimize settings with echo, even in the OR. For epicardial lead placement, it is particularly beneficial to utilize echo in the OR to minimize dyssynchrony before lead position is finalized.

Jamil Aboulhosen offered some guidance with pulmonary vasoactive medications in adults with a Fontan circuit. There has been resolution of PLE and normalization of mesenteric Doppler flow with sildenafil after Fontan. When the cardiac index and pulmonary blood flow increased with PDE 5i and sildenafil decreases respiratory rate and decreased minute ventilation at peak exercise. At the anaerobic threshold, subjects had significantly decreased ventilatory equivalents of CO₂, though with the addition of bosentan, peak VO₂, QOL, and BNP remained unchanged; however, NYHA class improved in 19%. The TEMPO trial with ERA's in Fontan circulation showed increased peak oxygen consumption and exercise time in the bosentan group. Exercise time increased, and there was immediate and short-term improvement in functional capacity.



Karen Stout and Luke Burchill moderate the opening session while Ariane Marelli speaks.



Mike Landzberg debating the future of Melody Valve use while George Liu, and Harsimran Singh look on.



Some work and some play - Craig Broberg leads the hike near Skamania Lodge.

Fontan Associated Liver Disease was presented by Dr. Curt Daniels reviewing discussions from the ACC Stakeholders meeting in 2015. There is clear evidence of progression over time, though difficult to predict. There are multiple risk scores including the VAST Score, the Liver Damage Score, the MELD XI and Child's Pugh. The proposed mechanism of injury is stretch-related as the mechanism of injury with micro-thrombosis. There is a suggestion that inspiratory muscle training may be beneficial in failing Fontan physiology.

Candace Silversides presented a discussion on pregnancy, aortopathy, and the risks of aortic dissection. Many women entered into a pregnancy with a prior history of dissection. She offered that the 40mm cutoff for Marfan's may be overstated.

Twenty-seven percent of pregnancies occurred in women with aortas >40mm, and they had no dissection. Many women have no known aortopathy prior to presenting with their dissection. Remember that in Turner Syndrome aortic dissection can occur even with a normal root size. Dissection rate is approximately 2%. In the patients with Turner Syndrome, the Aortic-Sized Index (ASI) should be assessed as this compensates for short stature." In those patients with Vascular Ehlers Danlos, 12 of the 83 women undergoing over 180 pregnancies, died during pregnancy or early post-partum with the additional risk of uterine rupture of 5% per pregnancy. Beta blockers are still recommended for prevention of aortic complications during pregnancy though there is no data.

Yoav Dori from CHOP/HUP offered some novel considerations for management of PLE in single-ventricle Fontans. Ninety percent of the patients after Fontan have nontraumatic chylo thorax. Five types of Plastic Bronchitis (PB) have been defined; 75% of PB with CHD cessation of casting with management. They are approaching 98% procedural success. They have identified liver lymphatic flow disorders. Patients with recurrent ascites, but without PLE, may be a candidate for lymphatic interventions.

Joe Kay discussed the use of the Melody Valve and monitoring for endocarditis, stent fractures, etc. Melody valve 7 year outcomes are now available, and re-intervention is often required within 6 years. There is an increasing risk of stent fracture over time in the IDE trial. The Melody valve may contribute to aortic root compression, deformation, and even erosion. There is something different about the Contegra valves that predispose them to endocarditis, and life-long aspirin is recommended, as well as SBE prophylaxis.

Heidi Connolly presented excellent data and experience regarding the fate of the Bioprosthetic Pulmonary Valve. She specifically discussed a normal Contegra conduit with normal hemodynamic Doppler for a pulmonary valve and the characteristics of 310 normal PVR in adults. Even normally functioning valves have a mean gradient of 15 mmHg. TTE and TEE are complementary techniques. Severe obstruction is a fairly common presentation for bacterial endocarditis. Twelve percent of bioprosthetic valves subsequently developed thrombosis. By Echo, the risk factors include an increasing gradient >50% of baseline, with restricted cusp mobility, thickened leaflets, and there is a recommendation to proceed to TEE when in doubt. In the setting of subclinical thrombosis, there is a risk of thrombotic obstruction of a Melody valve-in-valve. There is ongoing research to determine if 1-3 months of VKA prevent BPVT, and there is a certain residual risk with senescence – wear and tear phenomenon.

Adrienne Kovacs offered some excellent strategies for thriving and surviving as an ACHD practitioner. It is important to remember self-care for the practitioners who are providing the day-to-day care for our population.

Carole Warnes closed the meeting with a keynote address. She gave us a glimpse of Congenital Heart Disease past with a nod to one of the founders of Congenital Heart Surgery, Dr. Lillehei. There are an increasing number of ACHD patients, and many are having repeat operations - 5, 6 and 7 or more times.



The poster session was well-attended with many interesting presentations.

The population continues to age; now 65% of patients are >40 years old. We are also increasingly challenged by multi-factorial Fontan failure in an aging single-ventricle population.

The 27th International Symposium in Adult Congenital Heart Disease will be held September 14th-16th, 2017 in Cincinnati, OH.

CCT

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CONGENITAL CARDIOLOGY TODAY

© 2016 by Congenital Cardiology Today (ISSN 1554-7787-print; ISSN 1554-0499-online).
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Publication Company Address:

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