CONGENITAL CARDIOLOGY TODAY

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

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North American Edition

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Upcoming Medical Meetings
(See website for additional meetings)

BIT's 6th Annual International Congress of Cardiology (ICC-2014)
Nov. 13-16, 2014; Haiku, China
www.bitlifesciences.com/icc2014/program.asp

RSNA 2014 (Radiological Society of North America)
Nov. 30-Dec. 5, 2014; Chicago, IL USA
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PCICS 10th International Conference
Dec. 11-14, 2014; Miami, FL USA
www.pcics.org/meeting_detail.php?m_id=65

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Cardiac CTA Imaging with 3D Reconstruction in Diagnosis of a Unique Presentation of Ebstein's Anomaly in an Infant: A Case Report

By Melissa Symon, BS; Randy Richardson, MD

Introduction

Ebstein's Anomaly (EA) is a rare congenital heart defect (0.5% of all congenital heart defects[1]) in which there is apical displacement of the tricuspid valve (TV). The resulting right ventricle, distal to the displaced valve, is reduced in size. The remaining ventricle proximal to the valve becomes enlarged and atrialized, which in combination with the atrium results in a massively enlarged right atrium. In many cases of EA, the septal leaflets of the displaced tricuspid valve adhere to the ventricular wall(s). The resultant TV regurgitation further dilates the already abnormally increased volume of the right atrium. Tricuspid regurgitation, ventricular dysfunction, insufficient pulmonary blood flow, and arrhythmias contribute to symptoms of cyanosis and congestive heart failure.[2] The degree of right ventricular dysfunction depends on the size of the functioning ventricle and the severity of the tricuspid regurgitation.

EA is often found in association with additional congenital cardiac malformations, patent foramen ovale, atrial septal defects, pulmonary valve stenosis, ventricular septal defects, coarctation of the aorta, and transposition of the great arteries.[3,4] Malformations, when in combination, represent complex cardiac anatomy. The use of cardiac CT angiogram and perspective three-dimensional reconstruction with color-coding provides a useful clinical tool in such scenarios. We describe a case in which CTA angiography was employed in the diagnosis and improved visualization and comprehension of an infant presenting with a unique cardiac anomaly: EA, Levo-Transposition of the Great Arteries, and hypoplastic ascending aorta.

"EA is often found in association with additional congenital cardiac malformations, patent foramen ovale, atrial septal defects, pulmonary valve stenosis, ventricular septal defects, coarctation of the aorta, and transposition of the great arteries.[3,4]"

Case Report

The patient isa large-for-gestational age baby girl born at term by Cesarean section with vacuum assist to a G5P4 female with negative serologies.

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At birth the infant was noted to be mildly centrally cyanotic and was treated with blow-by oxygen. Cardiac exam revealed a grade III/VI continuous murmur. At four hours of age, with mild respiratory distress, the infant was transferred to St. Joseph's Medical Center and admitted to the newborn ICU for further management.

Initial imaging studies, including chest X-ray, echocardiogram and CTA with 3D image revealed a complex cardiac diagnosis of Ebstein's Anomaly of the tricuspid valve, L-transposition of the great vessels, moderate aortic insufficiency, hypoplastic aortic arch, patent ductus arteriosus and atrial septal defect. Within twenty-four hours of life, the infant became acidotic, bradycardic and began desaturating: by Day Two, the infant was intubated and transferred to the cardio-thoracic ICU. Further diagnostic studies revealed a normal female karyotype, a copy number gain of chromosome band 3p12.3 of approximately 0.424 Mbp in size, with no clear clinical relevance, and negative FISH performed with probe specific for the region deleted/duplicated in DiGeorge Syndrome/Velo-cardio-facial Syndrome. Electrocardiogram was abnormal demonstrating an intraventricular conduction delay, widened QRS with prolonged QT and right atrial enlargement.

On day fourteen the infant underwent her first cardiac surgery, a Damus-Kaye-Stansel procedure (anastamosis of proximal pulmonary artery to aorta bypassing systemic outflow obstruction) with arch reconstruction, tricuspid valve repair, and the placement of right ventricle to pulmonary artery homograph. In the operating room, the infant did not tolerate weaning from bypass, and the homograft was ligated and a Blalock-Taussig (systemic-to-pulmonary) shunt placed. Post-operative progression was unfavorable with the infant becoming increasingly hemodynamically unstable, and by day sixteen, the infant was initiated on extracorporeal membrane oxygenation (ECMO), placement of peritoneal dialysis catheter for management of acute renal failure, and lead revision of atrial pacemaker. The following day the infant returned to the operating room for revision of the tricuspid valve (Starnes procedure) and atrial reduction.

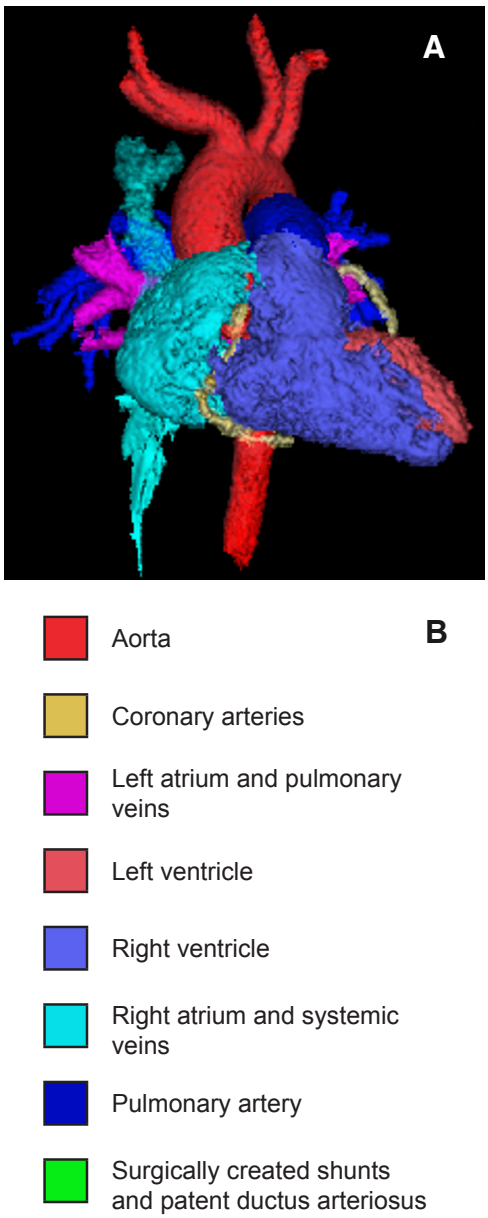

Figure 1: Normal cardiac anatomy (A) CTA demonstrating normal infant cardiac anatomy with standardized color coding; (B) Standardized color coding key: Aorta – red; Coronary Arteries – neutral; Left Atrium and Pulmonary Veins – pink; Left Ventricle – salmon; Right Ventricle – violet; Right Atrium and Systemic Veins – blue; Pulmonary Artery – dark blue; and, Surgically created shunts and Patent Ductus Arteriosus – green.

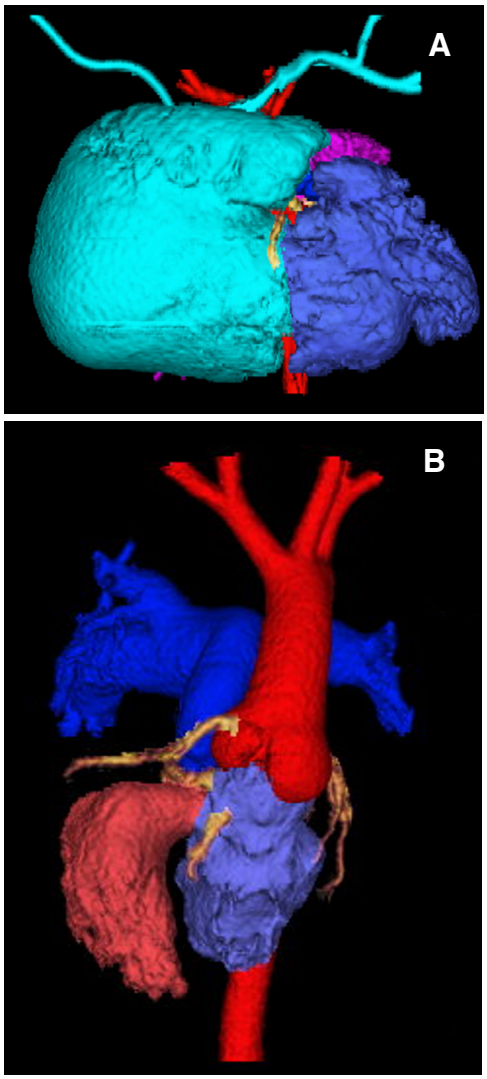

Figure 2. Examples of CTA of congenital cardiac malformations (A) Ebstein's Anomaly demonstrating apical displacement of tricuspid valve with resulting dilated Right Atrium (blue), and (B) L-Transposition of the Great Arteries with morphological Left Ventricle (salmon) on the anatomic right, and morphologic Right Ventricle (violet) in the anatomic left position. Note, atria are not shown.

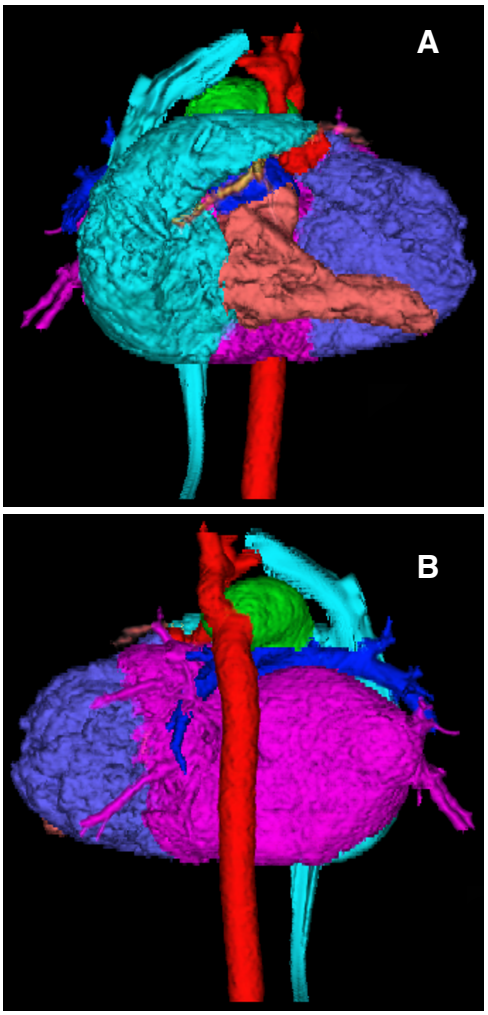

Figure 3. Cardiac findings in our infant presenting with respiratory distress. CTA demonstrates complex unique cardiac anatomy: Ebstein's Anomaly with massively dilated Left Atrium (pink), L-Transposition of the Great Arteries, hypoplastic ascending aorta (red), and Patent Ductus Arteriosus (green). (A) Anterior view. (B) Posterior view.

During the following two months, the hospital course was complicated by enterococcus bacteremia, omental artery bleed repaired by exploratory laparotomy, and tracheostomy placement for ventilator dependence. At age two-and-a-half months the infant was transferred to an inpatient facility for long-term care. Over the next month the infant was readmitted on several occasions for respiratory distress and hypoxia requiring increased ventilation. By age three months the decision was made to return to the operating room for bi-directional Glenn procedure (superior vena cave connected directly to the right pulmonary artery). Shortly thereafter, the infant again was placed on ECMO. The infant continued to deteriorate, developing focal seizures, bilateral hemothoraces, severe coagulopathy, refractory hypotension, and multisystem organ failure. At this point, it was decided to withdraw ECMO. The patient expired peacefully in her father's arms at age three-and-a-half months.

Cardiac CT Angiography Technique and 3D Reconstruction Methods[5]

Prospective computed tomography (CT) using intermittent scanning is an appropriate option for infant imaging due to the lower radiation exposure in comparison to retrospective CT in which scanning is continuous. Fast scanning times of approximately four to five seconds require short duration of anesthesia. Images generated are manipulated and reconstructed on external workstation by the radiologist. Anatomical components (aorta, pulmonary artery, ventricles, etc.) are segmented, and standardized color-coding applied to the three-dimensional image (Figure 1).

Cardiac function may be assessed by evaluation of ejection fraction and stroke volume as a function of end diastolic and systolic volumes.

CTA Findings

The cardiac findings are as follows: the left atrium is significantly enlarged with a large atrial septal defect. As typical with EA (Figure 2A), our case demonstrates an apically displaced dysplastic tricuspid valve adherent to the intra-ventricular septum in a morphological right ventrical (confirmed by the presence of a moderator band). Unusual to the typical presentation of EA is that the location of the tricuspid valve and the morphological right ventricle is located on the left-side of the heart, functioning as a left ventricle. This atrioventricular discordance in combination with a regurgitant left-sided (tricuspid) valve contributes to the massively dilated left atrium. Reflecting the discordance of the morphologic right ventricle to the left side of the heart is a morphological left ventricle, located on the right side. In addition, the outflow tracts, the aorta and pulmonary trunk are switched, consistent with a Levo-transposition of the great vessels (Figure 2B): the aorta arises from the morphologic right ventricle (located on the left), and the pulmonary trunk from the morphologic left ventricle (located on the right). The result of this double transposition of the ventricles and outflow tracts is a congenitally-corrected left-sided blood flow to the aorta and right-sided blood flow to the pulmonary trunk. The aorta itself is significantly hypoplastic, with a large patent ductus arteriosus measuring 13 mm x 24 mm (Figure 3).

Discussion

The severity spectrum of EA ranges from mild, in which the individual remains asymptomatic, to severe, which is often fatal in-utero.[6] Early clinical presentation correlates with a worse prognosis, with mortality

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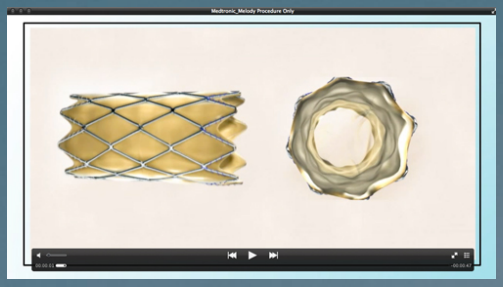

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risk decreasing eight-fold when diagnosed beyond the first year of life in comparison to earlier diagnosis.[7] Symptomatic neonates, as demonstrated by our case, represent a challenging group with poor outcomes and high mortality, and early intervention is recommended to optimize survival of these individuals. For those that fail to maintain adequate oxygen saturation with weaning of prostaglandins, or the development of congestive heart failure, surgery remains the recommended therapy.[8] Comparisons of shunts, valve repair valve replacement and biventricular repair, have been made in an attempt to guide surgical management of EA. [8,9,2] Procedure selection based on the specific anatomy and severity is recommended; however, guidelines are limited to variations of pure Ebstein malformations without additional defects. Increasingly complex cases in which more than one type of deformity co-exist, as in our case, warrants evaluation on an individual basis.

Cardiac CTA with three-dimensional reconstruction and color-coding, as demonstrated by our case, provides a modality through which it is possible to fully visualize unique cardiac anatomy as well as extra-cardiac structural relationships: specifically, coronary arteries, pulmonary arteries and pulmonary veins. Three-dimensional images viewed by clinicians allow for thorough understanding and accurate diagnosis, and may be of particular use in pre-surgical planning. Further enhancing understanding of complex anomalous cardiac anatomy, data from the 3D reconstructed images can be extracted, and with the utilization of 3D printers produce exact replica models which can further guide treatment and surgical approach. Three-dimensional imaging and models have been proven useful in improving the outcomes of oral and maxillofacial surgery.[10] Although cardiac surgery requires different techniques, and arguably surgical outcomes and successes are not evaluated in the same way as reconstruction surgery, knowledge of existing anatomy is no less important, and may be extremely beneficial to the surgeon performing these procedures. A better pre-procedural understanding of the anatomy may potentially decrease the length of surgery, and consequently, the duration of anesthesia.

The associated exposure to radiation with CT imaging is an important factor that must be addressed when considering utilization of this type of imaging study, especially in the case of pediatric patients. Pediatric patients present a complicated patient population in this respect as typically life expectancy is sufficient to warrant concerns of radiation-induced neoplasms. Radiation exposure of the CTA imaging study in our case has been estimated to be 29.63 DLP, which is the equivalent exposure of approximately six portable chest x-rays. MRI is a well-accepted alternate imaging modality that is often preferred; although more expensive, MRI has no associated radiation exposure. Unfortunately, MRI imaging of pediatric patients requires anesthesia, introducing an additional risk, which may be particularly concerning for unstable patients with complex congenital heart disease. As in the case of our patient, and other infants with severe cardiac anomalies, the benefit of accurate understanding of unique anatomy by the surgeon pre-procedurally, may significantly improve mortality outcomes, and therefore, the risk of radiation exposure may be heavily outweighed by the increased rate of survival. Further studies are required to evaluate the effect of use of 3-D CTA imaging on clinical outcomes and validate it as a clinically useful tool.

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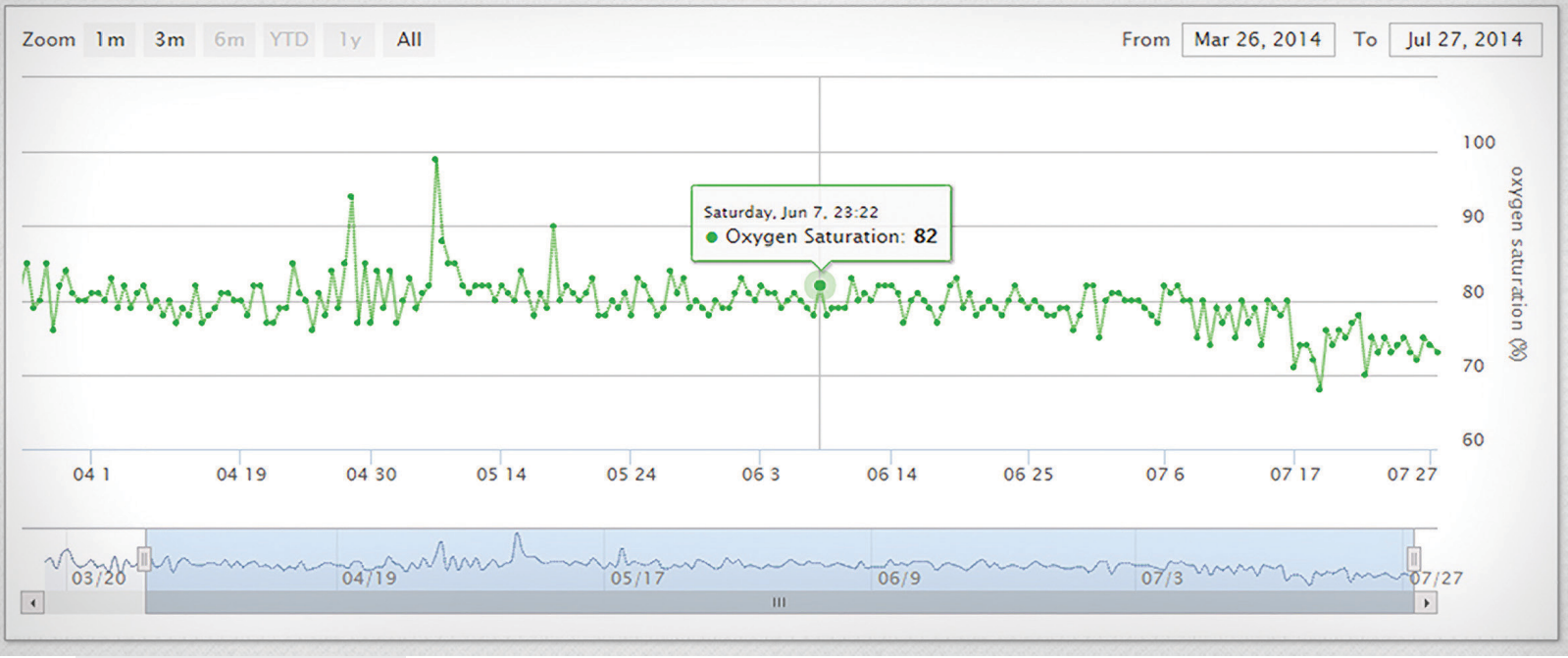

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"Further studies are required to evaluate the effect of use of 3-D CTA imaging on clinical outcomes and validate it as a clinically useful tool."

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Biographical Sketch of Corresponding Author

Melissa Symon is currently a fourth year medical student at Creighton University School of Medicine, Phoenix Regional Campus. Born and raised in the UK, she completed her Bachelor of Science in Biology at The University of Manchester, England. Melissa relocated to Seattle, Washington, where she worked in occupational and environmental health research at the University of Washington. It was during this time that she decided to pursue a career in medicine. She is currently applying for residency.

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BC/BE Pediatric Cardiac Electrophysiologist

The Carman and Ann Adams Department of Pediatrics at the Children's Hospital of Michigan, Wayne State University School of Medicine is recruiting a Board Certified/Board Eligible Pediatric Cardiac Electrophysiologist at the Assistant or Associate Professor level to join an established Electrophysiology program. IBHRE certification is strongly recommended but not required. The position will be available early 2015. Please circulate this to interested staff.

The successful applicant must be licensed/licensable to practice in the state of Michigan and will join the current senior Pediatric Electrophysiologist and dedicated EP nurse practitioner as well as 15 other cardiologists, 2 surgeons and 7 other mid-level providers in Detroit's largest cardiology and only Pediatric EP and cardiovascular surgical programs. The position includes providing invasive and non-invasive electrophysiology services, including inpatient and outpatient consultations, EP studies with 3D mapping and catheter ablations as well device implant and explant to both pediatric and adult congenital heart patients. Expertise in pacing/ICD lead extraction is strongly recommended. Some general cardiology duties as well as Resident/Fellow teaching are to be expected.

In addition to EP, the Division of Cardiology has established echocardiography, interventional, adult congenital, heart failure/transplant, pulmonary hypertension, as well as Cardiology Fellowship training programs. Drawing from a population of approximately 5 million people in Southeast Michigan, the division provides about 7500 outpatient visits, 700 cardiac catheterizations, and 110-130 EP/pacemaker procedures annually and is in the process of a major physical expansion. We currently actively follow 200 patients after pacemaker implantation and 55 patients with ICD's. Two congenital heart surgeons perform over 300 operations annually including heart transplant. Inpatient work is performed in Children's Hospital of Michigan, the only free-standing children's hospital in Michigan and the teaching hospital for Wayne State University. In addition, there are numerous opportunities for clinical, translational and basic science research. Salary will be commensurate with training and experience.

BC/BE Pediatric Cardiologist

The Carman and Ann Adams Department of Pediatrics at the Children's Hospital of Michigan, Wayne State University School of Medicine is recruiting a Board Certified/Board Eligible Pediatric Cardiologist with expertise in cardiac non-invasive imaging at the Assistant or Associate Professor level to join an established non-invasive imaging program. The position will be available early 2015. Please circulate this to interested staff.

The successful applicant must be licensed/licensable to practice in the state of Michigan and will join 15 other pediatric cardiologists. Drawing from a population of approximately 5 million people in Southeast Michigan, the division provides about 7500 outpatient visits, 9000+ echocardiograms, 700 cardiac catheterizations, and 200+ cardiac MRI procedures annually. There is an active telemedicine program interpreting 1300+ echocardiograms at outside institutions. Two congenital heart surgeons perform over 300 operations annually including heart transplant. Inpatient work is performed in Children's Hospital of Michigan, the only free-standing children's hospital in Michigan and the teaching hospital for Wayne State University. In addition, there are numerous opportunities for clinical, translational and basic science research. Salary will be commensurate with training and experience.

The primary assignment will involve inpatient and outpatient consultations, performing/interpreting echocardiograms, TEE and fetal echocardiograms. Participation in the CMR service is dependent upon experience and interest. Some general cardiology duties as well as Resident/Fellow teaching are to be expected.

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The 7th National Conference of the Adult Congenital Heart Association – "Translating Quality of Life into Quality of Care" Meeting Review

By Tabitha G. Moe, MD

The *7th National Conference of the Adult Congenital Heart Association - "Translating Quality of Life into Quality of Care"* was held September 4-7th, 2014 at the Holiday Inn Chicago Mart Plaza. Over 500 attendees included practitioners, patients, families and advocates. Three separate tracks allowed for concurrent clinical and patient education. Dr. Curt Daniels presented the most recent criteria for accrediting adult congenital heart disease (ACHD) programs. There will be two levels: one a comprehensive program with full surgical and interventional cardiac services, and the other a community program with referrals to a larger center. The presentation was well-received as a challenge to all ACHD programs to raise the standards of care.

A whirlwind update on the care of single-ventricle patients followed. Dr. Brian Kogon, from Emory University, suggested that conversion to Fontan physiology perhaps should be delayed, as there may be a finite longevity to Fontan circulation. He also suggested utilizing the infrahepatic portion of the IVC to form the total cavopulmonary connection. Dr. Gruschen Veldtman from Cincinnati Children's discussed prophylactic pacemaker placement for Fontan palliation revision as therapy for expected sinus nodal dysfunction following surgery. The idea of mechanical support therapies, including LVAD, was raised by a number of speakers and destination support devices seem to be gaining momentum. Pulmonary vasodilator therapy in Fontans remains highly contested.

Team-based care was a theme throughout the conference. Sue Fernandes, LPD, PA-C, from Stanford presented the idea of Whole Person Care vis a vis Patient-Centered Care. This should include routine psychological and social worker support; two areas that are often under-utilized in clinical practice.

Dr. Alexander Opotowsky, from Boston Children's presented some collaborative research opportunities including the ACHD quality enhancement research initiative (QuERI), and the Eisenmenger QuERI. Both trials are currently active. Site enrollment information is available at clinicaltrials.gov.

An amazing overview of the extracardiac effects of congenital heart disease drew on the team-based concept with experts in Pulmonology, Hepatology, and Nephrology discussing multi-organ involvement. A few pearls: from Dr. Opotowsky, the forced vital capacity predicts exercise capacity and the average DLCO is 61% in Fontans. Monitoring patients for pulmonary hypertension is indicated as twenty percent of associated pulmonary hypertension is congenital heart disease. Eisenmenger patient's should be evaluated for iron deficiency, and should not be phlebotomized. Dr. Naser Ammash from the Mayo Clinic, discussed cardiovascular accidents and the role of anticoagulation in ACHD. Warfarin continues to be the gold standard, as the pharmacodynamics of the novel oral anticoagulants in Fontan physiology remain unclear. Dr. Michael Earing advocated for the use of Cystatin-C over serum creatinine in evaluating signs of early kidney injury and developing protocols specifically to minimize risks in ACHD.

Back left: Scott Klewer, MD; Middle row (left-to-right): Edward Rhee, MD; Chris Heller, Cindy Huie, Tabitha Moe, MD; Brett Cottle; Front row (left-to-right): Elissa Heller, Andi Young, Joann Cottle, PA

Dr. Michel Ilbawi offered perspectives on evaluating the dysfunctional right ventricle with echo-based tricuspid annular plane systolic excursion, presystolic tricuspid regurgitation, volume indices, and strain and strain rates, as well as myocardial isovolumic acceleration. He presented a glimpse into the future with possible tricuspid valve chordal relocation and valvular reapproximation.

Dr. Joseph Kay spoke about evaluating and treating baffles: leaks should be therapeutically anticoagulated with warfarin, baffle bare metal stents treated with aspirin for six months. He advocated for every newly evaluated atrial switch patient to undergo an agitated saline injection study with contrast as the gold standard for possible baffle leak.

The Gala was a star-studded affair with Glenn Tringali the new National Executive Director awarding the former director Amy Verstappen with a new award named in her honor. Amy is a triple threat: ACHD warrior, compassionate leader, and devoted advocate. The next meeting date has not been released as yet, but you can get updated information at: www.achaheart.org.

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ACHA conference faculty: left-to-right: Ali Zaidi, MD; Michael Earing, MD; Salil Ginde, MD; Ami Bhatt, MD; Naser M. Ammash; Richard Krasuski, MD; Alexander Opotowsky, MD; Fred Wu, MD; Jamil Aboulhosn, MD. Photo compliments of Robert Levy Photography (Rhlphoto.com).

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Outstanding Barth Syndrome Conference Energizes Patient-Family-Researcher Community

By Matthew J. Toth, PhD

Over 330 people attended the *7th International Scientific, Medical & Family Conference on Barth Syndrome*, held at the Hilton Clearwater Beach Resort in Clearwater, Florida on June 23-28th, 2014. The conference was hailed as an outstanding success by the individuals with Barth Syndrome, family members, clinicians and researchers who attended. More than half of the Scientific and Medical (SciMed) attendees indicated this was their first Barth Syndrome Foundation (BSF) Conference and more than 95% stated they would attend a future conference.

An Essential Gathering to Facilitate Research and Healthcare

The attendees included 47 Barth Syndrome individuals (cardiomyopathy, neutropenia, growth delay, muscle weakness, and early fatigue) and their families. Also, 75 researchers, physicians, or healthcare professionals (most of the experts who work on this rare X-linked mitochondrial disease), were also there--the most ever.

The scientific and medical program of the conference included 22 speakers covering topic areas such as: Mitochondrial Lipids, Clinical Studies on Barth Syndrome, Mitochondrial Physiology, and Animal Models. There were 25 posters on display from which four poster presenters were selected to speak about their work. These international conferences are sponsored by BSF and its affiliates, while the scientific and medical sessions were supported in part by a R13 grant from NHLBI, NCATS, and NICHD.

Highly Commended Dual-Track Conference Format

BSF conferences are held over two days. As in previous years, there were separate programs formulated for families or for scientists/physicians. Nevertheless, both groups shared times together including mealtimes, the Clinical Studies session, the poster session, and the Friday night social event, which made for an enjoyable, educational, and inspiring event for everyone.

In the days that proceeded the SciMed sessions, five IRB-approved clinical studies were performed to take advantage of this large assembly of Barth Syndrome individuals. The Family sessions provided educational sessions with many researchers, healthcare workers, and physicians leading the talks and discussions.

New for 2014 were meetings where Barth Syndrome individuals discussed how they cope with their medical condition. Carrier sisters and mothers also discussed the many issues which they face. Both of these topics led BSF for the first time to advertise "Requests For Applications" (RFAs) as part of BSF's Research Grant Program.

Conference Keynote Lecture

Barry J. Byrne, MD, PhD, delivered the Keynote Lecture on Orphan Product Development in the Era of Personalized Medicine. Dr. Byrne is a renowned pediatric cardiologist, Director of the Powell Gene Therapy Center at the University of Florida, and a major champion for finding treatments for individuals with rare diseases. He has been a particularly close friend of BSF over the years.

The Scientific/Medical Presentations — Thursday

The morning sessions focused on mitochondrial lipids concentrating on the unique lipid associated with Barth Syndrome and with mitochondria —cardiolipin. A new understanding of the physiological roles for this unusual lipid was discussed which included exploring specific therapeutic ideas.

The afternoon session focused on clinical studies, including particularly encouraging reports from the two clinics specializing in Barth Syndrome care.

Bristol Clinic Lessons and Bezafibrate Treatment Trial

Dr. Colin Steward of the Bristol, England clinic (operational since 2010), which is supported by the National Health Service of the United Kingdom, provided insightful lessons. Prominent among these suggestions were the importance of multidisciplinary teams working among healthcare workers involved in the care of Barth Syndrome individuals, and avoidance wherever possible of the tendency to "subcontract" medical care to specialists working in different areas. In a disease that affects so many systems, this can result in affected boys/men having unmanageable numbers of consultations, with negative consequences for their schooling or employment. In addition, there is a definite benefit in having the older boys/young men with Barth Syndrome interact with their younger peers in clinic and non-clinic situations.

Importantly for the BSF community, Dr. Steward and colleagues have received preliminary support from the UK's National Institute for Health Research to clinically test whether bezafibrate treatment may be therapeutic in Barth Syndrome. Ann Bowron, FRCPath, reported on a subgroup of Barth syndrome individuals whose cardiolipin dysfunction levels are intermediate between the biochemical values of the unaffected population and of other Barth syndrome individuals. This unique Bristol subgroup appears to be less affected than others with this mitochondrial disease. Details of this intermediate phenotype have now been published as an Open Access article available via BSF's website (Bowron et al., J. Inherit. Metab. Dis., August 12, 2014).

"The scientific and medical program of the conference included 22 speakers covering topic areas such as: Mitochondrial Lipids, Clinical Studies on Barth Syndrome, Mitochondrial Physiology, and Animal Models."

Kennedy Krieger Institute Clinic

Dr. Hilary Vernon reported on the clinic established for the last two years at the Kennedy Krieger Institute in Baltimore, MD. Dr. Vernon presented and discussed the biochemical profiles of Barth Syndrome individuals which showed: increased 3-methylglutaconic acid in plasma; increased urine levels of organic acids; lower pre-albumin, arginine, and cystine levels in plasma; higher levels of tyrosine, proline, and asparagine in plasma; and unremarkable cholesterol levels. None of these individual biochemical values correlated with age or with each other or with neutrophil counts (neutropenia being a common symptom of Barth Syndrome). In addition, the Baltimore clinic also performed an analysis of the female carriers of Barth Syndrome (asymptomatic), but they could find no biochemical abnormalities.

Physician group shot.

Further Studies in France and USA

Dr. Jean Donadieu of the Trousseau University Hospital in Paris, updated the group about his efforts to gain recognition of Barth Syndrome in France, assisted by the BSF affiliate, Association Barth France. Dr. Donadieu and colleagues have published a retrospective analysis of the French experience with Barth Syndrome which is available as an Open Access publication available on BSF's website (Rigaud, et al., Orphanet J. Rare Dis. 2013).

Dr. Todd Cade of Washington University in St. Louis reported on his metabolic studies supported in large part by his R01 grant. In preliminary data with stable and radioactive tracers, under resting, exercising, and recovery periods, it appears that Barth Syndrome individuals show lower palmitate oxidation, increased protein breakdown (but not oxidation), increased glucose uptake (but not oxidation), and increased lactate production.

Dr. Stacey Reynolds (University of Florida) spoke about her investigations into the taste and feeding behaviors of Barth Syndrome individuals. Dr. Reynolds confirmed the salty-cheesy food preference in Barth syndrome individuals and their different taste sensitivities and eating habits. Dr. Reynolds has discovered a high incidence of super-tasters for PTC (bitter) among Barth Syndrome individuals, but no super-tasters were found for sodium benzoate (salty).

Dr. John Jefferies (Cincinnati Children's Medical Center) described in detail pediatric cardiac care and how different cardiomyopathies require different care plans. Dr. Jefferies emphasized that Barth Syndrome individuals should be screened and have continuous monitoring for ECG changes, arrhythmias, and cardiac dysfunction to avoid bad outcomes, and that appropriate medical and device-based therapies should be provided for the appropriate patients.

The Scientific/Medical Presentations — Friday The morning sessions covered mitochondrial physiology. Dr. William Pu (Boston Children's Hospital) presented his recent Nature Medicine article (also available as Open Access on BSF's website) using iPS cells differentiated into cardiomyocytes to overtly show the beating dysfunction of Barth Syndrome and to analyze, in great detail, what is altered biochemically. This heart-on-a-chip technology has profound implications for basic and applied research beyond Barth Syndrome.

The afternoon session discussed animal models of Barth syndrome and specifically the work done in several laboratories with the tafazzin knockdown mouse model of Barth Syndrome. Of particular interest was the report about a potential knockout mouse model from Dr. Douglas Strathdee (Beatson Institute for Cancer Research, Glasgow), and a report from Dr. Michael Chin (University of Washington) using enzyme replacement therapy with the knockdown mouse model of Barth Syndrome.

2014 Varner Award for Pioneers in Science and Medicine

The Varner Award, presented to the pioneers of the Science and Medicine of Barth

Meeting conference attendees in the shape of the BSF logo.

syndrome, was presented to Iris Gonzalez, PhD of A. I. DuPont Hospital for Children in Wilmington, DE. Dr. Gonzalez has been an integral part of BSF and its international Scientific and Medical Advisory Board from the very beginning. Dr. Gonzalez personally catalogs all the human tafazzin gene mutations —tafazzin is the gene that when mutated causes Barth Syndrome. Dr. Gonzalez's insights into tafazzin gene structure and function contribute to our basic knowledge of this disease, but she never forgets that behind these mutations are real people suffering real problems. Dr. Gonzalez graciously remarked during her acceptance speech that the Varner Award was her Nobel Prize!

Final Observations of a Stimulating Conference

There was a large amount of unpublished information revealed at the 2014 Meeting which is eagerly anticipated appearing in the literature. Not everyone can be mentioned in the space of this short article. Please accept apologies and grateful thanks to everyone who contributed to this successful conference.

Survey feedback from the attendees was extremely positive—the best ever for these conferences. For 2016, we will continue to encourage young and new researchers to attend the meeting, continue to showcase the therapeutic and potentially therapeutic activities that are taking place, and make more of an effort to attract physicians, who may be caring for a single Barth Syndrome individual, to attend. Videos of many of the speakers' presentations are available on BSF'S website (www.barthsyndrome.org) for anyone to access and review.

CCT

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Medical News, Products & Information

By Tony Carlson, Senior Editor, CCT

Effective Method to Prevent and Treat Heart Failure

Researchers have discovered a previously unknown cardiac molecule that could provide a key to treating, and preventing, heart failure.

The newly discovered molecule provides the heart with a tool to block a protein that orchestrates genetic disruptions when the heart is subjected to stress, such as high blood pressure.

When the research team, led by Ching-Pin Chang, MD, PhD, Associate Professor of Medicine at the Indiana University School of Medicine, restored levels of the newly discovered molecule in mice experiencing heart failure, the progression to heart failure was stopped. The research was published in the online edition of the journal *Nature*.

The newly discovered molecule is known as a long non-coding RNA. RNA's usual role is to carry instructions -- the code -- from the DNA in a cell's nucleus to the machinery in the cell that produces proteins necessary for cell activities. In recent years, scientists have discovered several types of RNA that are not involved in protein coding but act on their own. The role in the heart of long non-coding RNA has been unknown.

But the researchers determined that the newly discovered non-coding RNA, which they named "Myheart" -- for myosin heavy-chain-associated RNA transcript -- is responsible for controlling a protein called BRG1 (pronounced "berg-1"). In earlier research published in *Nature* in 2010, Dr. Chang and his colleagues discovered that BRG1 plays a crucial role in the development of the heart in the fetus.

But as the heart grows and needs to mature into its adult form, BRG1 is no longer needed, so very little of it is produced. That is, until the adult heart is subjected to significant stress such as high blood pressure or damage from a heart attack. Dr. Chang's previous research showed that in those conditions, BRG1 re-emerges and begins altering the heart's genetic activity, leading to heart failure. At the same time, production of Myheart is suppressed, so BRG1 can latch onto the DNA and alter the genetic material unchecked.

In the current Nature paper, the researchers reported that in mice with stress-induced high levels of BRG1, they were able to restore Myheart to normal levels using gene transfer technology. Restoring Myheart levels blocked BRG1 actions and prevented heart failure, they said.

"I think of Myheart as a molecular crowbar that pries BRG1 off the genomic DNA and prevents it from manipulating genetic activity," said Dr. Chang, Director of Molecular and Translational Medicine at the Krannert Institute of Cardiology.

Although the results in mice would suggest testing Myheart against heart failure in humans, it is too large -- by molecular standards -- to be delivered as a drug, Dr. Chang said.

So he and his colleagues now are working to identify smaller portions of the Myheart molecule that are key to its ability to block BRG1. Such a subsection of the Myheart molecule could lead to a compound to test in human trials.

In addition to Dr. Chang and Pei Han, PhD, first author of the paper, investigators contributing to the research included Wei Li, Jin Yang and Peng-Sheng Chen of the IU School of Medicine; Chiou-Hong Lin, Ching Shang, Sylvia T. Nuernberg, Kevin Kai Jin, Weihong Xu, Chieh-Yu Lin, Chien-Jung Lin, Yiqin Xiong, Huan-Chieh Chien, Euan Ashley, Daniel Bernstein and Thomas Quertermous of the Stanford University School of Medicine; Bin Zhou of the Albert Einstein College of Medicine; and Huei-Sheng Vincent Chen of the Sanford/Burnham Medical Research Institute.

The research was supported by the American Heart Association (AHA; Established Investigator Award 12EIA8960018); the National Institutes of Health (NIH; HL118087, HL121197, HL109512, HL105194, HL78931, HL71140, HL116997, HL111770); California Institute of Regenerative Medicine (CIRM; RN2-00909, RB2-01512, RB4-06276); Stanford Heart Center Research Program; the IU School of Medicine-IU Health Strategic Research Initiative; the IU Physician-Scientist Initiative, endowed by Lilly Endowment; the Lucile Packard Foundation for Children's Health; the March of Dimes Foundation (#6-FY11-260); the Oak Foundation; and the Baxter Foundation.

Henry Ford Hospital Replaces Heart Valve Outside the Heart

Newswise — For the first time in the United States, doctors at Henry Ford Hospital used a minimally invasive procedure to replace a failing, hard-to-reach heart valve with a new one – and placed it just outside the heart.

Due to prior medical procedures, the metro Detroit woman was not a candidate for traditional open heart surgery to replace her failing tricuspid valve. One major U.S. medical center turned her down for treatment. Doctors at another said her only option was a heart transplant.

Henry Ford is the first hospital in the U.S. to perform the unique, transcatheter tricuspid valve replacement, which was pioneered in Germany. The woman's tricuspid valve, one of four valves that regulates blood flow in the heart, was replaced during the 2-hour procedure July 31st. The woman stayed five days in Henry Ford Hospital and was released to her home.

"There are a lot of people who have damage to the tricuspid valve, and the surgery is risky, so doctors just try to give them medical therapy," says William O'Neill, MD, Medical Director of Henry Ford's Center for Structural Heart Disease and lead physician for the procedure. "They get a lot of swelling and severe liver congestion. They're in and out of the hospital, and it really causes a lot of morbitity. So there's a huge, unmet clinical need. Individuals with this type of valve problem now have another option."

Approximately 5 million people in the U.S. are diagnosed with heart valve disease annually. With an aging population that is often too frail for open-heart surgery, more than 20,000 Americans die of the disease each year, according to the American Heart Association.

More than 9,100 heart patients undergo tricuspid valve surgery in the United States annually, according to the Society of Thoracic Surgeons. Replacing a tricuspid valve is one of the more difficult heart surgeries due to the valve's location, typically requiring open heart surgery, Dr. O'Neill explains. The tricuspid valve is in the middle of the heart, in between the right ventricle and the right atrium.

In the unique procedure at Henry Ford Hospital, Dr. O'Neill threaded a catheter through a vein in the patient's groin to her upper abdomen. There, he inserted the TAVR valve at the junction of the right atrium and the inferior vena cava (IVC), the main vein that brings deoxygenated blood back into the heart.

The Henry Ford team first braced the inside of the IVC with a metal, expandable stent. He then used the catheter to insert and expand a TAVR valve to fit snugly inside.

The team used 3D modeling to create a working replica of the patient's heart, which helped them properly plan the procedure and choose an appropriately sized valve in advance.

Once deployed, the new valve stopped blood from leaking and pooling in the patient's abdomen and lower extremities.

JANUARY MEETING FOCUS

Sport Cardiology & Sudden Cardiac Arrest in the Young

Jan. 23-24, 2015
Disney Grand Californian Resort, Anaheim, CA USA
www.choc.org/conference

Overview
An opportunity for all those involved in the care of athletes and young individuals in general to gain up-to-date education relating to the effects of athletic training, and the diagnosis and management of individuals with inherited cardiac diseases, in the hope of preventing tragic young sudden deaths in the future.

This conference draws together experts in the fields of sports cardiology, inherited cardiac diseases and sudden cardiac death in the young, who will present contemporary topics and developments in their respective fields.

Major Topics Covered
- Workshop: Interpretation of the ECG in the Young Athlete
- Genomics of SCA
- Channelopathies, Exercise and SCD
- Risk Stratification of SCA - Debates
- "Real World" Cases in Sports Cardiology – Case Based Discussions
- "50 Shades of Grey" Zone Athlete: Distinguishing Normal Adaptation from Pathology
- Sports Cardiology in Youth Athletes
- Basic Exercise Physiology and Sports Cardiology
- Unknown Cases from Audience
- Panel discussion /Audience Q&A
- Plus more...

Program Director:
Anjan S. Batra, MD, FHRS, Director of Electrophysiology
Children's Hospital of Orange County
Section Chief and Vice Chair of Pediatrics
University of California, Irvine

Registration Options:
Online: https://www.regonline.com/Register/Checkin.aspx?EventID=1600685
Early Bird Registration (until 12/19/14):
Physicians - $400; Fellows/Nursing - $300; Residents/Medical Students - $25

Regular Registration after 12/19/14:
Physicians - $450; Fellows/Nursing - $350; Residents/Medical Students - $75

CHOC designates this live activity for a maximum of 12.0 CME hours of AMA PRA Category 1 Credits™.

CHiP NETWORK
CONGENITAL HEART PROFESSIONALS

WHAT IS THE CHiP NETWORK? - The CHiP Network, the Congenital Heart Professionals Network, is designed to provide a single global list of all CHD-interested professionals in order to:
- Connect pediatric and adult CHD-interested professionals to events, conferences, research opportunities and employment
- Keep members up with the literature through the monthly *Journal Watch* service
- Increase education and provider awareness of new developments
- Bring the pediatric and adult congenital heart communities into closer contact
- Offer a communication tool for critical issues

WHO SHOULD PARTICIPATE? - The CHIP Network is all inclusive and is comprised of everyone who considers themselves a congenital heart professional or administrator, including:
- Pediatric cardiologists
- ACHD cardiologists
- RNs and APNs
- Cardiac surgeons
- Cardiac care associates
- Trainees/fellows
- Administrators
- Psychologists and mental health professionals.
- Researchers/scientists
- Intensivists
- Anesthetists
- Industry representatives

OUR SUPPORTING PARTNERS:
- Adult Congenital Heart Association
- Asia Pacific Society for ACHD
- Children's Hospital of Philadelphia Cardiology meeting
- Cincinnati Children's Hospital
- Congenital Cardiology Today (official publication of the CHiP Network)
- Congenital Heart Surgeons Society
- International Society for Adult Congenital Heart Disease
- Japanese Society of ACHD
- Johns Hopkins All Children's Heart Institute
- North American ACHD program
- Paediatric Cardiac Society of South Africa
- Pan Arab Congenital Heart Disease Association
- PCICS
- PICS
- Specialty Review in Pediatric Cardiology
- World Congress of Pediatric Cardiology and Cardiac Surgery

JOIN US - Membership is Free!
The CHiP Network management committee invites the participation of other organizations who want to communicate with all or some of the congenital heart professionals on this list. Please contact Dr. Gary Webb (gary.webb@cchmc.org) to ask that your organization's or institution's name be added to the list of partner organizations.

HOW TO REGISTER
Register at www.chipnetwork.org. It takes only a minute and you can unsubscribe at any time.

Cincinnati Children's Heart Institute

Funded by Cincinnati Children's Heart Institute

The Barth Syndrome Foundation

Barth Syndrome Foundation

P.O. Box 974, Perry, FL 32348
Tel: 850.223.1128 info@barthsyndrome.org www.barthsyndrome.org
Symptoms: Cardiomyopathy, Neutropenia, Muscle Weakness, Exercise Intolerance, Growth Retardation

"There's already a huge drop in the pressure in the abdomen," Dr. O'Neill says. Doctors monitored pressure through catheters inserted in the IVC above and below the new valve.

Dr. O'Neill was assisted by Adam Greenbaum, MD, Director of the Cardiac Catheterization Lab at Henry Ford Hospital, and visiting cardiologist Brian O'Neill, MD, Assistant Professor of Medicine at the Temple Heart and Vascular Center.

For more information visit www.henryford.com/structuralheart.

Toshiba America Medical Systems, Inc: 2014 RSNA Showcase

Toshiba America Medical Systems, Inc. will demonstrate the following diagnostic imaging technologies at this year's *Radiological Society of North America* (RSNA) annual meeting, November 30-December 5th at McCormick Place in Chicago.

CT:

Toshiba's CT Solutions Provide Safer Exams for Aquilion ONE and Aquilion PRIME Families

As today's healthcare providers strive to improve the quality of care they offer while decreasing costs, Toshiba continues to develop technology that aligns with these goals. Toshiba has made software and hardware enhancements (some pending 510(k) clearance) to its Aquilion CT scanners as well as new dynamic clinical applications that take advantage of wide detector CT technology, to improve clinicians' ability to accurately plan a patient's treatment.

Toshiba Showcases New Patient-Friendly PET/CT System

To advance the care clinicians provide, Toshiba's Celesteion PET/CT system delivers a more comfortable patient experience with the industry's largest bore, widest field-of-view, dose reduction technology and fastest imaging. The versatile system combines high-performance PET and CT for all radiation and oncology imaging needs, including tumor detection, treatment evaluation and CT simulation. As healthcare reform puts a greater emphasis on patient satisfaction, the Celesteion's numerous patient-friendly features are designed for this new landscape.

Skin Cells Can Be Engineered Into Pulmonary Valves for Pediatric Patients - New Valves May Grow with Patients and May Have Lower Rejection Rates

Newswise — Chicago – Researchers have found a way to take a pediatric patient's skin cells, reprogram the skin cells to function as heart valvular cells, and then use the cells as part of a tissue-engineered pulmonary valve. A proof of concept study in the September 2014 issue of The *Annals of Thoracic Surgery* provides more detail on this scientific development.

"Current valve replacements cannot grow with patients as they age, but the use of a patient-specific pulmonary valve would introduce a 'living' valvular construct that should grow with the patient. Our study is particularly important for pediatric patients who often require repeated operations for pulmonary valve replacements," said lead author David L. Simpson, PhD, from the University of Maryland School of Medicine in Baltimore.

Dr. Simpson, senior co-author Sunjay Kaushal, MD, PhD, and colleagues designed a process to transform skin cells from a simple biopsy into cells that become an important ingredient in a tissue-engineered pulmonary valve.

The pulmonary valve is a crescent-shaped valve that lies between the heart's right ventricle and pulmonary artery. It is responsible for moving blood from the heart into the lungs.

While the study was conducted in vitro (outside of the body), the next step will be implanting the new valves into patients to test their durability and longevity.

"We created a pulmonary valve that is unique to the individual patient and contains living cells from that patient. That valve is less likely to be destroyed by the patient's immune system, thus improving the outcome and hopefully increasing the quality of life for our patient," said Dr. Kaushal. "In the future, it may be possible to generate this pulmonary valve by using a blood sample instead of a skin biopsy."

Dr. Simpson added that he hopes the study will encourage additional research in tissue engineering and entice more people to enter the field, "Hopefully, growing interest and research in this field will translate more quickly into clinical application."

It is estimated that nearly 800 patients per year could potentially benefit from bioengineered patient-specific pulmonary valves, according to data from the STS Congenital Heart Surgery Database. The Database, which collects information from more than 95% of hospitals in the US and Canada that perform pediatric and congenital heart surgery, shows that approximately 3,200 patients underwent pulmonary valve replacement during a 4-year period from January 2010 to December 2013.

The Simpson study was a collaboration between the University of Maryland School of Medicine in Baltimore and the Northwestern University Feinberg School of Medicine in Chicago. Additional co-authors include: Brody Wehman, MD; Yekaterina Galat; Sudhish Sharma, PhD; Rachana Mishra, PhD; and Vasiliy Galat, PhD (senior co-author).

CitationsAnn Thorac Surg 2014;98:947–54.

Diary Assists Cardiac Patients and Their Health Care Teams in Documenting Vital Information to Keep Them Well

In June of 2013, Raven Marie Dietrich Deel unexpectedly contracted a viral infection of the heart (myocarditis) and a bacterial infection of the heart (endocarditis), causing permanent damage and congestive heart failure.

Now, one year later, Raven Marie is able to maintain wonderfully without a mechanical heart thanks to her careful documentation of everything she eats, drinks, and expels. With the urging of her doctors, she has published the format she created to organize this vital information. The result, *Cardiac Vitals Journal*, allows other heart patients to also benefit from the aid of a diary for heart patients.

This useful and comprehensive diary, notes Raven Marie, is an easy go-to when doctors, nutritionists, and physical therapists have questions.

Cardiac Vitals Journal is designed to make it easy to document daily medications, food choices, sodium, fluid intake, fluid output, potassium, calories, carbohydrates, fiber, protein, cholesterol, sugar, diuretic, a.m. and p.m. blood pressure and heart rates, insulin and blood sugar detailing for those who need it, supplements, and daily exercise habits.

Readers can also transfer information from this first section of the book to a section called

"Information for My Doctor" that can be torn out and taken with them to appointments.

An additional section titled "Questions, Notes, & Progress Reports" allows readers to organize questions they might think of that they want to remember to ask their healthcare providers.

She comments, "I have been able to maintain without a mechanical heart because I documented all the information I noticed my health care team documenting in every hospital I was in throughout the country. There is no other diary out there for cardiac patients that provides for this information. My doctors loved the idea, and that is why I wrote the book. It's great for you, your family members, and your home health providers."

Author: Raven Marie Dietrich Deel, author of *Cardiac Vitals Journal* and the poetry/short story book, *Walking through Misty-Colored Memories,* unexpectedly ended up with endocarditis (bacterial infection of the heart) and myocarditis (viral infection of the heart) in June of 2013, which destroyed the left ventricle and affected the mitral valve of her heart (her ejection fraction rate, or heart function, is currently under 20%). She felt she could fulfill a need in the heart failure community with this diary.

Seattle Children's Research Institute Awarded $1.6 Million to Lead National Study on Heart Defects - *New drug intervention could improve outcomes for infants with congenital conditions*

Newswise — The U.S. Food and Drug Administration's Office of Orphan Products Development has awarded Seattle Children's Research Institute a $1.6 million grant to lead a 4-year, multi-site clinical trial aimed at improving long-term health in infants born with a heart defect.

Dr. Michael Portman, Director of Pediatric Cardiovascular Research at Seattle Children's Hospital and an investigator in Seattle Children's Research Institute's Center for Developmental Therapeutics, will be investigating whether treating infants on cardiopulmonary bypass with thyroid hormone supplement may help them recover more quickly and with fewer side effects.

"The societal cost of congenital heart disease is huge," Portman said. "These are children who suffer lifelong effects. If we can use a medication that's already proven to be safe to help them recover from heart surgery then that could have a major impact on their lives."

The trial will begin enrolling participants next month and will include patients from Seattle Children's, Stanford University Lucille Packard Children's Hospital and Los Angeles Children's Hospital.

Portman previously discovered the benefit of thyroid supplementation for children on bypass in a study published in 2010 in the American Heart Association's academic journal, *Circulation*.

Knowing thyroid hormone levels decrease in infants and children while on cardiopulmonary bypass, Portman led a team of researchers to test the safety of thyroid hormone supplementation in infants and children after cardiac surgery to determine whether it might improve patient outcomes.

The randomized clinical trial, which included children under 2 years old, revealed that giving patients thyroid hormone was safe for all ages and improved clinical outcomes in patients under 5 months old. A second

study looking specifically at patients under 5 months of age is required to modify the labeling of thyroid hormone supplement, which would impact the current standard of care.

"I anticipate this study will confirm the results of our previous research so that clinicians may begin prescribing this supplement to those patients that would benefit most," Portman said.

Congenital heart disease occurs in approximately 32,000 infants per year and approximately 38% of these patients will have one or more surgical procedures in their lifetime. Over the past 20 years, more and more have undergone cardiopulmonary bypass. While the surgical procedures themselves have improved, little research has been done to investigate whether medications might be beneficial to patients after surgery. Infants are especially likely to spend prolonged time on mechanical ventilation due to poor heart function after surgery and fluid build-up in the lungs.

Portman believes these effects are worsened because cardiopulmonary bypass causes an inflammatory response that suppresses normal thyroid hormone production. Still, relatively few randomized clinical trials have evaluated agents, like thyroid supplement, provided to these infants after cardiopulmonary bypass.

"Seattle Children's is focused on using cutting-edge research to offer the best in clinical care," Portman said. "If this study is successful, patients will spend less time on a ventilator, less time in cardiac intensive care, and less time in the hospital. They will have the chance to grow up and live longer, healthier lives."

Rady Children's to Establish Pediatric Genomics and Systems Medicine Institute

Newswise — Rady Children's Hospital-San Diego has taken a major step forward in the research of childhood diseases with the establishment of the Rady Pediatric Genomics and Systems Medicine Institute at Rady Children's. Ernest Rady and family have made yet another extraordinary investment in the health and well-being of children by pledging $120 million to support pediatric research and innovation at the Institute. The gift is the largest donation ever made to Rady Children's Hospital Foundation. Rady Children's Board of Trustees has committed an additional $40 million in funding to support the Institute.

"After more than one and one half years of evaluating the design and mission of an Institute at Rady Children's, this Institute will assemble a team of world class scientists, researchers and clinicians who will focus their talents on preventing, diagnosing, treating and curing childhood disease through genomics and systems medicine research," said David F. Hale, Chairman of the Board. "The Institute will work closely with UC San Diego and establish relationships with other academic and research institutions, companies involved in genomics research and other children's hospitals to advance the mission of the Institute," said Hale.

"The commitment Ernest Rady and the Board has made is truly transformative," said Donald Kearns, MD, President of Rady Children's. "This Institute and gift will secure Rady Children's place at the leading edge of research, discovery and innovation into childhood disease and injury, and set national and international standards for pediatric care."

"As we move into an exciting new era of medicine it is our responsibility to encourage the ambitious research and innovation that will accelerate the process by which discoveries are made and translated into cures," said Mr. Rady.

Discoveries in genomics and the resulting emergence of personalized medicine hold unprecedented promise, yet the methods and resources for translation into cures and treatments are still to be identified. This gift will allow Rady Children's to invest these funds in the highest and most current needs to keep pace with new technology and stay on the cutting edge of genomics and systems medicine. It is planned that the Institute will be housed in two facilities, 7910 Frost Street, adjacent to Rady Children's, and a location in the Torrey Pines area, situated among other distinguished research institutes.

Mr. Rady served as Chair of the Children's Hospital and Health Center Board from 1990 to 1993, during which time the Board oversaw construction of the hospital's Rose Pavilion. He also served as a member of that same Board, and as a member of the Children's Hospital-San Diego Board, in the 1980s and '90s. During that time, Mr. Rady and his wife, Evelyn, made several major donations to Children's; the main entrance lobby and the second floor Medical/Surgical Center of the Rose Pavilion are named for Dr. Max and Rose Rady, a tribute to Mr. Rady's parents. In 2006, in recognition of a $60 million gift by the Rady family and American Assets, Inc., the company Mr. Rady founded in 1967, the Board of Trustees voted unanimously to rename the hospital Rady Children's Hospital-San Diego.

"Not only will generations of children benefit from Mr. Rady's extraordinary legacy of giving, our entire region is the better for it," said Paul Hering, Chair, Rady Children's Foundation Board. "Very few people in the world have succeeded in making such a positive, lasting philanthropic impact on our children now and in the future."

For more information, visit www.rchsd.org.

CONGENITAL CARDIOLOGY TODAY

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