

# CONGENITAL CARDIOLOGY TODAY

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**SCAI 2014 Scientific Sessions**  
May 28, 2014; Las Vegas, NV USA  
[www.scai.org](http://www.scai.org)

**Basic & Advanced Fetal Cardiology Symposium Workshop**  
Jun 5-6, 2014; Chicago, IL USA  
<http://fetalcardiacsymposium.com/>

**PICS-AICS**  
(includes: Cardiac Imaging for Structural Heart Disease - A Special One Day Symposium at PICS)  
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## Ruptured Non-Coronary Sinus of Valsalva Aneurysm Causing Right Heart Failure

By Nicole Brown, MD; Gruschen Veldtman, MBChB, FRCP; David Luis Simón Morales, MD; Christopher Learn, MD

### Introduction

Sinus of Valsalva Aneurysms (SVAs) are rare cardiac defects that can be either congenital or acquired. The greatest concern is the risk of rupture. Even if unruptured, they can obstruct adjacent structures such as the coronary artery ostium or the right ventricular outflow tract. SVA are also associated with malignant arrhythmias and endocarditis,<sup>1</sup> and there is 3:1 male predominance. Although still rare, SVAs are both more common and up to five times more likely to rupture among Asian populations compared to Western populations.<sup>2,3</sup> Rupture most commonly occurs in patients between 20-30 years of age.<sup>1</sup>

The aortic sinuses are named after Antonio Valsalva, an Italian anatomist and physician. They extend from the aortic annulus to the sinotubular junction. The function of the sinuses of Valsalva remains incompletely understood and a topic of ongoing research. They provide space between the open aortic valve leaflets and the coronary ostia. The vortices present in the sinuses are thought to facilitate coronary blood flow and reduce stress on the aortic leaflets during closure.<sup>4,5</sup> Comparative models of different aortic grafts showed higher stress in the cylindrical model

compared to the bulbous sinus design.<sup>4</sup> Preservation or reconstruction of the sinuses during valve-sparing aortic root surgery leads to re-creation of the vortices in the sinuses. This is believed to support normal leaflet movement, coaptation height, and valve durability. Despite abnormal leaflet motion seen with root reimplantation without aortic sinus re-creation, there is no definitive proof of adverse effects on valve durability.<sup>5</sup>

Abnormalities of sinus anatomy, such as the presence of an aneurysm, may profoundly alter the distribution of forces at the base of the aorta and disrupt aortic valve function. These altered forces may in turn worsen inherent medial weakness within an already compromised sinus. We report the case of a young man with a known non-coronary sinus of Valsalva aneurysm who presented following rupture of the aneurysm into the right atrium (RA).

***"Sinus of Valsalva Aneurysms (SVAs) are rare cardiac defects that can be either congenital or acquired. The greatest concern is the risk of rupture."***



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## Case Report

A 26-year-old male with partial trisomy 15 and severe global developmental delay suffered a right-sided embolic stroke at 14 years of age. This led to a diagnosis of a non-coronary sinus of Valsalva aneurysm. Surgical intervention was not pursued because of his multiple developmental and medical issues, including severe scoliosis and a non-functional dysplastic right kidney. He was instead started on warfarin. He was followed for over 10 years by pediatric cardiology with no new symptoms, a relatively stable international normalized ratio (INR), and no significant change by echocardiography.

At the time of his initial visit with the Cincinnati Children's Adolescent and Adult Congenital Heart Disease (AACHD) team, he was clinically stable with no cardiovascular symptoms, no murmurs, and stable electrocardiogram (ECG) findings. His echocardiogram showed a large non-coronary sinus of Valsalva aneurysm with stable dimensions (3.34 x 1.17 cm) that protruded into the right atrium at the level of the right ventricular inlet. In contrast to prior studies, an 8 mmHg mean inflow gradient across the tricuspid valve was noted. Right atrial dimensions remained normal despite the gradient. Elective repair and the risk of rupture were discussed with the parents, who opted for continued conservative management.

Nine months later, he presented to an outside emergency department with parental concerns of several weeks of changes in behavior, worsening oral intake, dry cough, and gradual onset of lower extremity edema. Doppler interrogation was negative for deep vein thrombosis. Labs revealed elevated blood urea nitrogen (BUN), uncharacteristically high INR, slightly elevated liver transaminases, mild thrombocytopenia, and proteinuria with hypoalbuminemia. No precipitating cause was found. He was re-evaluated by his primary care physician several days later. A renal ultrasound showed stable kidney findings, but new ascites and dilated hepatic veins. He was transferred directly to the AACHD clinic for further evaluation.

Vital signs were as follows: weight 46.3 kg (baseline 38.6 kg), blood pressure 113/71 mmHg, heart rate 100 bpm, respiratory rate 20 per minute, saturation 98% on room air. The exam showed very thin habitus, mild lethargy and injected sclera. His scoliosis, pectus excavatum, left-sided hemiplegia and contractures, and developmental delay were unchanged. His lungs were clear with normal work of breathing.

Cardiac exam revealed striking changes compared to his baseline exam. His precordium was now hyperdynamic with a right ventricular lift. On auscultation, he had a regular tachycardia, normal S1 and S2, a prominent S3/S4 gallop at the left lower sternal border, and a new harsh III/VI continuous murmur, loudest at the right upper sternal border with radiation throughout the precordium. Pulses were 2+ and symmetric with no radio-femoral delay, and capillary refill was normal with warm, well-perfused extremities. Mild 1+ edema was present in his lower extremities (left, slightly worse than right). Abdominal exam revealed ascites and an enlarged, pulsatile liver.

ECG showed normal sinus rhythm with right atrial enlargement and non-specific intra-ventricular conduction delay, and mildly prolonged QTc (Figure 1).

An echocardiogram confirmed rupture of the SVA through several small fenestrated defects of the non-coronary sinus with blood shunting left-to-right into the right atrium. Peak velocity across the defect was 4-5 m/sec. The aneurysm had increased in size (3.1cm x 2.8cm) and had an oblong shape. It protruded into the superior and medial right atrium and caused severe obstruction of RV inflow (Figure 2). The tricuspid valve mean gradient had increased to 14-15 mmHg,



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## OPPORTUNITY IN PEDIATRIC CARDIOLOGY MCALLEN, TEXAS

Driscoll Children's Hospital is advancing a comprehensive Heart Center to meet the healthcare needs of congenital heart patients in South Texas. The Center is recruiting a physician to support outpatient clinic activities in McAllen, TX and the Rio Grande Valley. Sub-specialty board eligible or certification is required. Spanish speaking is preferred.

Pediatric Cardiology has been an integral part of Driscoll Children's Hospital since 1962. The Hospital and the Heart Center are committed to bringing state-of-the-art technology and quality service to 31 counties in South Texas. In 2013, the Heart Center saw 9,500 outpatient and satellite visits; 6,121 echocardiograms, including 500 fetal echos, and 192 heart catheterizations (82% interventional). The Heart Center employees 8 physicians including 1 Electrophysiologist, 2 Interventional cardiologists, 1 MRI Imaging cardiologist, and 1 fetal cardiologist. Three pediatric cardio-thoracic surgeons deliver all aspects of surgical service including hybrid procedures.

The McAllen Clinic is a major clinic affiliated with Driscoll Children's Hospital. The cardiologist in McAllen will share a 1:4 call rotation involving consultation without post cardiovascular surgical care. Physicians will see challenging, complex patients in a beautiful, well-staffed clinic with 2 sonographers and in-house laboratory and radiology. The qualified physician will enjoy a young, fast growing patient base and a new University of Texas affiliated medical school.

McAllen and the Rio Grande Valley offer a vibrant, multicultural population. With the mild weather, it is a haven for year-round outdoor activities including golf, cycling, tennis and water sports and is 45 minutes from beautiful beaches. South Texas offers world class hunting, fishing, sailing and wind surfing. The cost of living in south Texas is low, and there is no state income tax.

### Contact Information

**Annette Shook, Executive Director,  
Physician Relations and Recruitment**  
Office: (361) 694 6807; cp: 361 877 7259  
email: [annette.shook@dchstx.org](mailto:annette.shook@dchstx.org)

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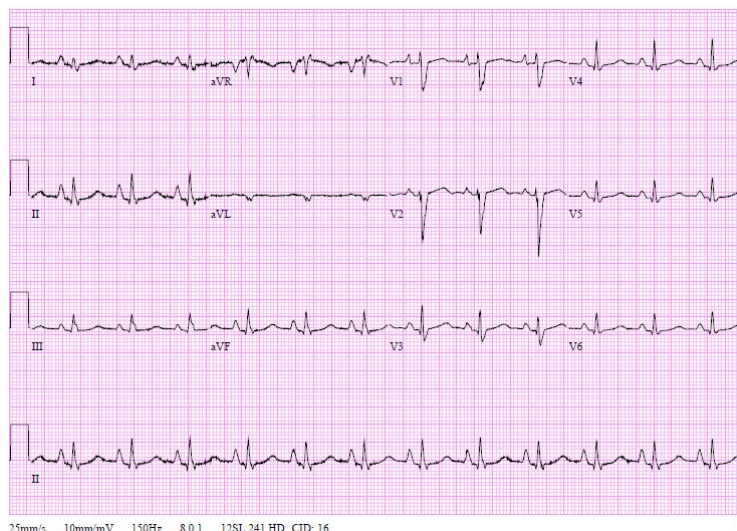


Figure 1. ECG showing sinus rhythm, right atrial enlargement, non-specific intraventricular conduction delay, and mildly prolonged QTc.

which may have been at least partially related to increased flow across the valve. There was no evidence of thrombus within the aneurysm. The aortic valve demonstrated only trivial regurgitation and no stenosis. The right atrium and ventricle were both dilated with prominent flow reversal in the hepatic veins. Biventricular systolic function was normal. A small-moderate pericardial effusion and abdominal ascites were also noted.

The patient was transferred to the cardiac intensive care unit and taken to the operating room the following morning. Routine pre-operative transesophageal echocardiogram (TEE) (Figure 3) confirmed findings on the transthoracic echocardiogram (TTE). Multiple fenestrations were seen in the non-coronary leaflet near the commissure with the right coronary leaflet. One jet was directed between the anterior and septal tricuspid valve leaflets and another larger jet was directed toward the right atrial appendage. The surgeon encountered a large effusion upon entering the pericardium. The right coronary artery was incidentally found to arise from the left sinus of Valsalva. There was a large amount of aneurysmal tissue present in the right atrium with adherence to the septal leaflet of the tricuspid valve. This caused poor tricuspid valve leaflet coaptation between the anterior and septal leaflets.

The surgeon resected some of the aneurysmal tissue within the RA. A trimmed patch of CorMatrix was used to close the defect from the aortic side, and the aneurysm tissue was also closed from the RA side to achieve a two-level closure. The aortic valve was repaired by reattaching some of the leaflet to the aortic wall, and it functioned well. The tricuspid valve was repaired by placing an Alfieri stitch between the anterior and septal leaflets.

Post-operative imaging showed no residual shunting and good function of the tricuspid and aortic valves. The patient did very well after surgery and had good spontaneous diuresis. He was discharged on the sixth post-operative day. We were able to stop his warfarin since the aneurysm and source of his prior embolic stroke

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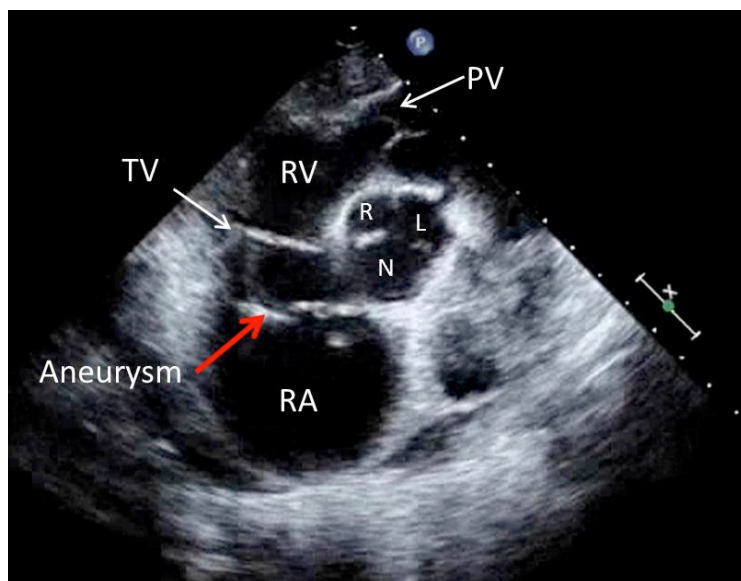


Figure 2a. Transthoracic echocardiogram: 2D parasternal short-axis view at the level of the aortic valve showing an aneurysm originating from the non-coronary sinus of Valsalva and protruding into the right atrium at the level of the tricuspid valve. RA-right atrium, RV-right ventricle, TV-tricuspid valve, PV-pulmonary valve, R, L, & N-right, left, and non-coronary aortic cusps.

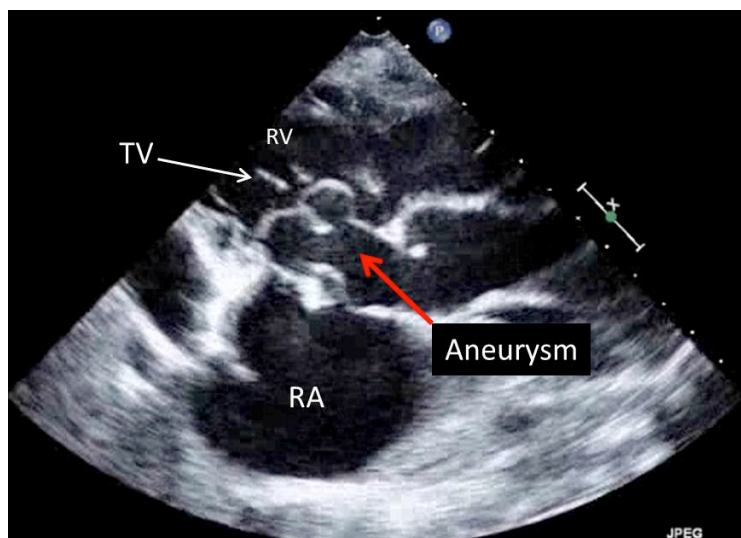


Figure 2b. Transthoracic echocardiogram: 2D modified parasternal long-axis view angled toward the tricuspid valve showing aneurysm of the non-coronary sinus of Valsalva obstructing right ventricular inflow. RA-right atrium, RV-right ventricle, TV-tricuspid valve.

had been removed. He was continued on aspirin for stroke prophylaxis. Within two weeks of his repair, he was back to his dry weight and had resolution of all signs of right heart failure.

## Discussion

This case represents a subacute presentation of a ruptured non-coronary SVA. About 50-70% of patients have a gradual onset of fatigue, dyspnea, and other symptoms of congestive heart failure.<sup>1</sup> Similarly, our patient was asymptomatic until after his aneurysm ruptured. He developed evidence of right heart failure over the course of about four weeks. This was secondary to both the left-to-right shunt from the aorta to the RA and the right ventricular inflow obstruction due to aneurysmal tissue from the SVA.

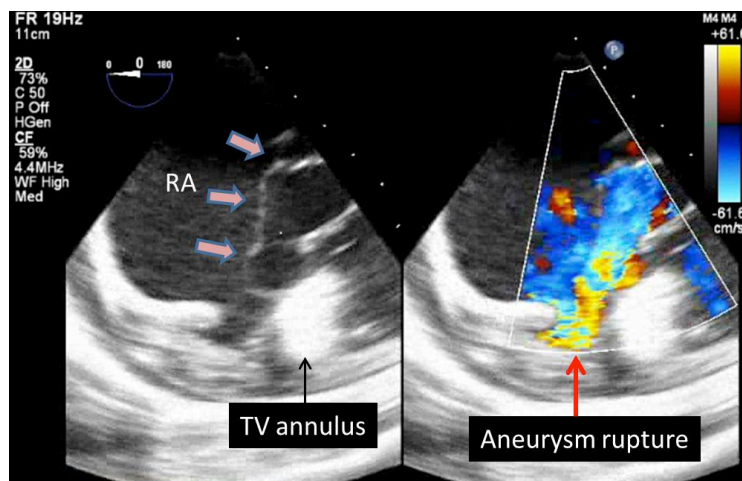


Figure 3a. Transesophageal echocardiogram: color compare image showing an enlarged right atrium, aneurysmal tissue superior to the tricuspid valve annulus, and color Doppler indicating high-velocity shunting from the aneurysm toward the right atrial appendage. RA-right atrium, wide arrows indicate non-coronary sinus of Valsalva aneurysm.

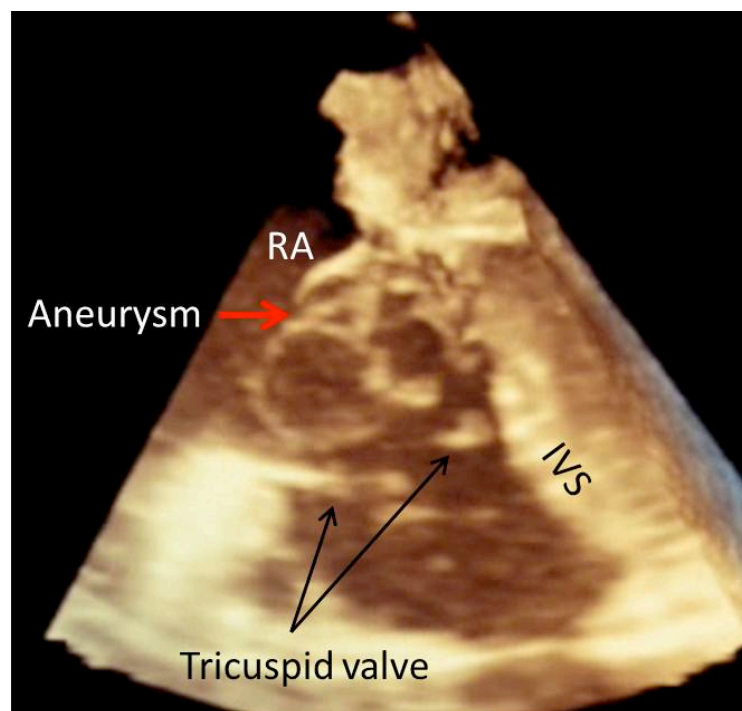


Figure 3b. Transesophageal echocardiogram: 3D image showing aneurysmal aortic sinus tissue extending across most of the right ventricular inlet. RA-right atrium, IVS-interventricular septum.

Aneurysms from the right sinus of Valsalva are most common (65-85%) with the majority of the rest originating from the noncoronary sinus (10-30%). An aneurysm from the left sinus occurs in less than 5% of cases.<sup>1,7</sup> SVA rupture is usually into a right heart chamber—more frequently into the RV for right SVA and into the RA for non-coronary SVA, but either can occur. Rarely, rupture occurs into the left ventricle, left atrium, pulmonary artery, or pericardium.<sup>1</sup>

Congenital aneurysms occur more frequently than acquired aneurysms, but are still rare among congenital lesions. Only 0.14-1.5 percent of congenital heart repairs are for congenital SVA.<sup>2</sup> They are thought to be due to abnormal elastic media between the aortic sinus and the aortic annulus and lack of continuity with the annulus fibrosis.<sup>2,6</sup> Syndromes associated with SVA include Klippel Feil, Turner, Marfan, Ehlers-Danlos, and Loays-Dietz. Trisomies 13 and 15, osteogenesis imperfecta, and





## Interventional Cardiologist Specializing in CHD

The Heart Center (THC) at Nationwide Children's Hospital (NCH), pediatric teaching facility for The Ohio State University in Columbus Ohio, is recruiting a Director of Interventional Cardiology. The applicant must have advanced fellowship training in complex transcatheter therapies in CHD. In addition, the applicant should have achieved the level of Director, Co-Director, or Associate Director of Interventional Cardiology at a free-standing Children's Hospital with a dedicated cardiac catheterization service. The applicant must be Board Certified in Pediatric Cardiology and have attained at least a level of Assistant or Associate Professor. Past experience as a P.I. or Co-P.I. for FDA sponsored device trials is required. In addition, the individual must have experience in all currently available transcatheter therapies for CHD, as well as a proven academic record in 1st authored scientific publications in high impact medical journals. The applicant must have demonstrated the ability to lead multi-center device trials and have a strong presence in being an invited speaker to national and international interventional scientific sessions. The ability to have been chosen as a program director of a scientific meeting is desirable, but not required. Being an expert in advanced imaging in the modern day cardiac catheterization suite also is desirable, along with experience in all forms of Hybrid therapies. A strong background in research, including animal studies, device design and development, is also required.

THC at NCH has been perennially rated in the top 10 Cardiology & Heart Surgery Centers in the USN&W Report, while NCH is also rated in the top 10 Children's Hospital in the US. THC consists of a unique organizational structure where all services; including cardiology, CT surgery, cardiac anesthesiology, cardiac intensive care, nursing, CV & Pulmonary Research center, and all support staff, are under 1 service line with 3 Co-Directors directly reporting to NCH administration leadership. The Center for CV & Pulmonary Research has a T-32 training grant that allows unique possibilities for advanced national & international training.

The Interventional Cardiology team participates in over 700 cardiac catheterizations with nearly 70% being interventional, and over 180 EP procedures, along with over 450 open-heart surgeries/year. There are 2 dedicated Hybrid Biplane FPD Cardiac Catheterization Suites that are adjacent to 2 Hybrid Cardiac Operative Suites and integrated into a state-of-the-art Cardiac Telemedicine Conference Center. Because of the innovative Hybrid program in THC, the applicant must be well skilled in these procedures, including exit angiography and interventions. All advanced transcatheter procedures are performed in THC, including all forms of transcatheter valve therapies...including Early Feasibility Trial for Native TPV replacement. A new, state-of-the-art CV Research Facility just opened with a Hybrid Cardiac Catheterization Suite and 5 bay Cardiac Surgical Suite. The applicant should be experienced and well trained to utilize this facility in order to design and test new, innovative medical devices and techniques.

THC at NCH, along with the OSU Wexner Medical Center is the home of 1 of the largest ACHD programs in the US, where over 3,500 patients are seen each year. Nearly 1/3 of all cardiac catheterizations are performed in ACHD patients, but some are performed at OSU (an additional 180 per year). Therefore, the applicant must have a strong background in performing complex transcatheter therapies in ACHD patients. In addition, select members of our team are also valuable members of the SHD team at OSU, serving as P.I. for TAVR procedures, mitral valve therapies, and will be involved in future innovative SHD therapies.

This position will require a unique skill set and wide experience in treating complex CHD in the fetus, newborn, child, and young and older adult with CHD. A unique opportunity exists to be part of a multidisciplinary team of dedicated healthcare professionals where innovation is the expectation and best outcomes the norm.

### Interested candidates are encouraged to submit their curriculum vitae to:

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rheumatoid arthritis have also been associated. We presume that our patient's SVA was congenital and likely due to his Partial Trisomy 15.

Acquired SVA has been attributed to bacterial endocarditis, syphilis, tuberculosis, atherosclerosis, and cystic medial necrosis. Trauma and hematoma as a complication of aortic valve surgery have also been linked to aneurysm formation.<sup>2,7</sup> Several classification schemes of SVA have been proposed. The earliest was by Sakakibara and Konno in 1962, and later by Ring, Guo, and most recently Xin-Jin et al.<sup>2,3,8</sup> No one classification system is widely used.

The patient's laboratory derangements were non-specific and could have been attributed to viral illness, injury of his single kidney, or poor nutrition. The red flags in this case were the loud continuous murmur and prominent gallop rhythm. These findings represented a definite change from his baseline cardiac exam and were harbingers of a serious cardiovascular problem. In this case, he was already known to have a SVA extending into the RA, thus rupture had to be a chief consideration. A new cardiac murmur, especially if continuous and accompanied by signs of right heart failure, should prompt concern for a ruptured SVA in any patient, and this clinical concern should guide appropriate echocardiogram image acquisition.

Historically, cardiac catheterization with aortography was the "gold standard" for diagnosing SVA; however, advances in echocardiography have allowed this non-invasive modality to be the initial diagnostic test of choice. Echocardiography can delineate the origin of the SVA, aneurysm size, protrusion into adjacent cardiac structures, and rupture, including shunt gradient. Aortic stenosis or regurgitation, tricuspid valve obstruction or insufficiency, chamber sizes, ventricular function, and pericardial effusion can also be investigated by echocardiography.<sup>1,9</sup>

Echocardiogram can detect anomalies associated with SVA as well. Ventricular septal defects are the most commonly associated anomaly. Some reports indicate a higher incidence with right sinus of Valsalva involvement. Aortic valve regurgitation is the second most common association, especially if rupture has occurred. In that case, up to 50 percent may require aortic valve surgery.<sup>1,10</sup> Other less common associations include other congenital cardiac anomalies, such as pulmonary stenosis, atrial septal defect, bicuspid aortic valve, Tetralogy of Fallot, patent ductus arteriosus, coarctation of the aorta, and subaortic stenosis.<sup>1</sup>

TTE is often sufficient for proceeding with surgical planning when the imaging is congruent with the clinical presentation and exam. It is also convenient and safe in more critically ill patients. If TTE is not sufficient, then TEE is likely to be valuable for defining the anatomy, sites of rupture, and valve involvement. Diagnostic cardiac catheterization is now reserved for stable patients in whom coronaries need to be better visualized.<sup>1,9</sup>

Untreated ruptured SVAs have demonstrated a mean survival period of one to four years in various series; therefore, early surgical intervention is needed. Congestive heart failure is the primary cause of death, but endocarditis may play a role in about 8% of patients. Indications for surgery include a ruptured SVA with left-to-right shunting, even in asymptomatic individuals. Urgent surgical intervention should be undertaken in any patient with signs of congestive heart failure. The indications for intervening in the case of an asymptomatic patient with an unruptured aneurysm are less clear. Increasing aneurysm size, evidence

of RV inflow or outflow tract obstruction, coronary compression, new symptoms, or presence of an underlying connective tissue disease should prompt consideration of surgery.<sup>1,10</sup>

The goal of surgical intervention is to repair the ruptured SVA, excise the aneurysmal tissue, and repair any associated defects without interrupting atrioventricular nodal conduction or causing valvular incompetence. There are no clinical trials indicating that any one surgical technique is superior.<sup>3</sup> Overall, surgical outcomes for ruptured SVA and symptomatic unruptured SVA are excellent with low morbidity and mortality in most series. Patients should continue to be monitored for recurrence of aneurysm formation, late aortic or tricuspid valve insufficiency, and residual septal defects.<sup>10</sup>

Transcatheter closure of ruptured SVAs with a septal occluder or similar device has been reported. While a non-operative approach may have several potential advantages, the long-term safety of such an approach is unknown. Surgery remains the gold standard of intervention for ruptured SVAs.<sup>11,12</sup>

In summary, SVAs are rare but potentially life-threatening defects that are usually congenital in origin. Even large aneurysms may be silent until rupture or obstruction occurs, most likely in the second or third decade of life. A harsh continuous murmur, especially if new and accompanied by signs of congestive heart failure, should trigger concern for ruptured SVA, regardless of whether the patient has a known history of aneurysm. Transthoracic echocardiogram is the diagnostic imaging modality of choice. Surgical repair of ruptured SVAs can typically be undertaken with good long-term outcomes.

Additional cases like the one above will be reviewed at *Congenital Heart Disease in the Adult: An International Symposium*, June 10-13, 2014 in Cincinnati. To register, visit [www.cincinnatichildrens.org/achdprogram](http://www.cincinnatichildrens.org/achdprogram).

#### Principal Author Biographical Sketch

Nicole Brown, MD completed residency in Internal Medicine and Pediatrics at the University of Cincinnati Medical Center and Cincinnati Children's Hospital Medical Center. She then completed a fellowship in Pediatric Cardiology at Children's Hospital of Pittsburgh of UPMC, where she also began her training in the care of adults with congenital heart disease. Dr. Brown is now participating in a two-year advanced fellowship in Adolescent and Adult Congenital Heart Disease at Cincinnati Children's Hospital Medical Center. She is board-certified in Internal Medicine and Pediatrics and board-eligible in Pediatric Cardiology. She also intends to sit for the Adult Congenital Heart Disease boards.

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**“Additional cases like the one above will be reviewed at Congenital Heart Disease in the Adult: An International Symposium, June 10-13, 2014 in Cincinnati. To register, visit [www.cincinnatichildrens.org/achdprogram](http://www.cincinnatichildrens.org/achdprogram).”**

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**CCT**



*Nicole Brown, MD  
Adolescent and Adult Congenital Heart  
Disease Fellow  
Heart Institute, Cincinnati Children's  
Hospital Medical Center  
3333 Burnet Ave.  
Cincinnati, OH 45229-3026 USA  
Phone: 513-803-2243  
Fax: 513-803-0079  
[nbrown1606@gmail.com](mailto:nbrown1606@gmail.com)*

#### **Corresponding Contributor**



*Gruschen Veldtman, MBChB, FRCP  
Director, Inpatient Adult Congenital Heart  
Disease Services  
Heart Institute, Cincinnati Children's  
Hospital Medical Center  
3333 Burnet Ave.  
Cincinnati, OH 45229-3026 USA  
Phone: 513-803-2243  
Fax: 513-803-0079  
[gruschen.veldtman@cchmc.org](mailto:gruschen.veldtman@cchmc.org)*



*David Luís Simón Morales, MD  
The Heart Institute  
Chief, Pediatric Cardiothoracic Surgery  
Professor of Surgery and of Pediatrics  
Clark-Helmsworth Chair of Pediatric  
Cardiothoracic Surgery  
Cincinnati Children's Hospital Medical  
Center  
The University of Cincinnati College of  
Medicine*



*Christopher Learn, MD  
Assistant Professor, University of  
Cincinnati Department of Pediatrics and  
Internal Medicine  
Heart Institute, Cincinnati Children's  
Hospital Medical Center  
Cincinnati, OH USA*

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# Placement of an Implantable Cardioverter Defibrillator in a Pediatric Patient with Heparin-Induced Thrombocytopenia with Thrombosis, Bridging with Argatroban

By Joshua A. Daily, MD; Cristina Tarango, MD; Richard J. Czosek, MD; Jeffrey B. Anderson, MD; David S. Spar, MD

## Case Report

A 17-year-old male (weight: 69 kg; body surface area: 1.27 m<sup>2</sup>) presented following cardiac arrest secondary to ventricular fibrillation. He was witnessed to collapse after running up and down a ramp. He was unresponsive and pulseless, and cardiopulmonary resuscitation was initiated. An automatic external defibrillator demonstrated that his rhythm was ventricular fibrillation. He received an initial shock of 200 Joules (J) which briefly converted him to sinus rhythm, but he reverted to ventricular fibrillation. He received a second shock of 300 J with conversion to sinus rhythm.

The patient developed significant respiratory failure and Acute Respiratory Distress Syndrome, and on hospital Day 5 was put on veno-venous Extracorporeal Membrane Oxygenation (ECMO) for 9 days. While on ECMO, he developed Heparin-Induced Thrombocytopenia with Thrombosis (HITT) secondary to the unfractionated heparin infusion with thrombocytopenia and thrombosis of his left cephalic and right basilic veins. The Heparin-PF4 antibody and 14C-serotonin release assay were positive consistent with a diagnosis of heparin-induced thrombocytopenia (HIT). The heparin was discontinued and argatroban, a direct thrombin inhibitor, was initiated. The thrombophilia work up was negative including normal protein C, normal protein S, normal antithrombin, negative Factor V Leiden, and negative prothrombin G20210A mutation. Full body ultrasound demonstrated no additional sources of thrombus.

He underwent an extensive cardiac work-up to determine the etiology of his ventricular fibrillation arrest. Cardiac CT and cardiac MRI were normal. His initial echocardiogram showed mildly depressed biventricular function with eventual normalization of function on subsequent echocardiograms.

We proceeded with diagnostic electrophysiology study and cardiac catheterization. Unfortunately, the post-ECMO course was complicated by chronic fevers which delayed these procedures. In the interim, he was transitioned to warfarin to both minimize the infection risk associated with a central line and to enable him to be discharged from the Intensive Care Unit (ICU); our institutional policy requires that patients on argatroban remain in the ICU. When the fevers resolved, we performed an electrophysiology study with ventricular stimulation protocol along with a hemodynamic cardiac catheterization with an endomyocardial biopsy all of which were normal. The warfarin was discontinued 5 days prior to the cardiac catheterization secondary to bleeding risk during arterial access so the patient was restarted on argatroban as a therapeutic bridge through the study.

We placed an implantable cardioverter defibrillator for secondary prevention approximately 30 days from the diagnosis of HITT. Since the patient had just transitioned back to argatroban for cardiac catheterization and had previously taken three weeks to become therapeutic on warfarin, the decision was made to use an argatroban bridge protocol for the ICD placement developed in conjunction with the hematology service (Table 1). The argatroban was initially dosed at 4 mcg/kg/min via continuous IV infusion. Argatroban was then stopped 6 hours prior to surgery. An international normalized ratio (INR), activated partial thromboplastin time (aPTT) (drawn 2 hours prior to surgery) and

activated clotting time (ACT) (drawn 1 hour prior to surgery) were verified and normalized prior to proceeding. The patient then underwent placement of a transvenous single chamber ICD in the left suprapectoral region without complication. Argatroban was then restarted at half the original dose, 2 mcg/kg/min continuous infusion 12 hours post-ICD placement. Warfarin was restarted 24 hours post-ICD placement after ensuring that there was no post-surgical hematoma formation. While the patient was on argatroban, PT, aPTT, INR, and diluted thrombin time were drawn every 6 hours and the infusion was titrated as needed for goal aPTTs 1.5-3 times baseline. Argatroban was continued until the warfarin was in a therapeutic range (goal chromogenic X activity of 17%-25%, INR was not used since it is also prolonged by Argatroban).

## Clinical Follow-Up Evaluation

The patient did not have any bleeding complications during his ICD placement, his remaining hospitalization, or 6-months post-ICD implant. He was discharged after warfarin reached therapeutic levels. Subsequent ultrasounds demonstrated complete resolution of all thrombi and the warfarin was discontinued after 4 months. The etiology of his cardiac arrest continues to remain unclear.

## Discussion

Pediatric patients requiring anticoagulation that necessitate placement of a Cardiac Implantable Electronic Device (CIED) represent a high-risk group. The primary complication of ICD placement is pocket hematoma formation which has an incidence in adults between 3.5-5% for those on warfarin or no anticoagulation<sup>1,2</sup> and greater than 20% for those bridged with Heparin.<sup>3,4</sup> For children, the risk of bleeding complication based upon type of anticoagulation is not well-defined, but in a review of a multicenter registry the overall incidence of hematoma was found to be 1.8%.<sup>5</sup> When deciding on an anticoagulation plan for ICD placement, we had to balance the risk of hematoma formation with the risk of

Table 1. Argatroban Protocol

Time Relative to Surgery	Action
	Argatroban 4 mcg/kg/min.
6 hours before	Discontinue Argatroban.
2 hours before	Verify that INR and aPTT have normalized.
1 hour before	Verify that ACT has normalized.
12 hours after	Restart Argatroban 2 mcg/kg/min and titrate for goal aPTT 1.5-3 times baseline (drawn every 6 hours).
24 hours after	Restart warfarin.
Indeterminate	Continue argatroban until chromogenic X activity is 17-25%.

INR international normalized ratio  
aPTT activated partial thromboplastin time  
ACT activated clotting time



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thromboembolic complications due to HIT. In adults with HIT, the incidence of thromboembolic complications is 38-76%<sup>6</sup>, often within 30 days of diagnosis, but it is significantly lower in those treated with Argatroban.<sup>7</sup> Pediatric data is much more limited, but overall the risk in children with and without treatment seems similar to adults.<sup>8</sup> In contrast to adults, though, HIT associated thrombosis in children is more often venous than arterial.<sup>9</sup>

Our review revealed no published experiences of anticoagulation in patients with HIT requiring ICD placement. In both children and adults with HIT and HITT, Argatroban has been used successfully to maintain patency in the cardiopulmonary bypass apparatus during cardiac surgery<sup>10</sup> and to prevent thrombosis during cardiac catheterization.<sup>11</sup> In addition, the safety, efficacy, and pharmacokinetics of Argatroban in children have been previously demonstrated,<sup>8</sup> and our institution's practice has been to use Argatroban in patients with HIT.

Our patient was receiving continuous anticoagulation with Argatroban at the time of his ICD placement. Rather than wait for him to become therapeutic on warfarin and significantly delay ICD implantation (previously therapeutic levels were reached in 3 weeks), the team decided to proceed with an Argatroban bridge. Given the high risk of hematoma formation in combination with the data suggesting that the risk for thrombus formation of short-term interruption of anticoagulation is very low, even in high thromboembolic risk patients<sup>12</sup>, the decision was made to hold his Argatroban for the case. The rate of pocket hematoma formation in patients bridged with heparin has been demonstrated to be the same whether heparin was restarted 6 hours or 24 hours after surgery, therefore, we chose to restart the Argatroban 12 hours after surgery.<sup>4</sup>

This report illustrates the unique challenges in treating patients at high thromboembolic risk who require a CIED. Though the patient tolerated the argatroban bridging protocol, this being a single reported case is a limitation of the protocol. This relatively uncommon occurrence of HIT in a patient requiring urgent device implantation will likely preclude extensive study on this population, so we chose to present this case and our argatroban protocol for other institutions that may face a similar clinical dilemma.

**Disclosures/ Conflicts of Interest:** none

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**CCT**

#### Corresponding Contributor



**Joshua A. Daily, MD**  
Cincinnati Children's Hospital Medical Center  
Division of Pediatric Cardiology  
The Heart Institute  
3333 Burnet Avenue MLC 2003  
Cincinnati, OH 45229, USA  
Ph: (513)636-4888  
Fax: 513-636-7996  
joshua.daily@cchmc.org



**Cristina Tarango, MD**  
Cincinnati Children's Hospital Medical Center  
Division of Hematology  
Cancer and Blood Diseases Institute  
Cincinnati, OH, USA



**Richard J. Czosek, MD**  
Cincinnati Children's Hospital Medical Center  
Division of Pediatric Cardiology  
The Heart Institute  
3333 Burnet Ave. MLC  
Cincinnati, OH 45229, USA



**Jeffrey B. Anderson, MD, MPH**  
Cincinnati Children's Hospital Medical Center  
Division of Pediatric Cardiology  
The Heart Institute  
3333 Burnet Ave. MLC  
Cincinnati, OH 45229, USA



**David S. Spar, MD**  
Cincinnati Children's Hospital Medical Center  
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# Does Renal Denervation Have a Future? TRENDS Conference Gives a Resounding “Yes”

By Sameer Gafoor, MD

The recent *TRENDS* conference in Frankfurt discussed at length the current status and future of renal denervation. Device-Based ....and Systemic Interventions was the focus of the recent *TRENDS* conference held in Frankfurt, Germany, to discuss at length the current status and future of renal denervation. Recent trial data has called the future of renal denervation into question, and this was discussed extensively.

## Friday, February 7, 2014

This started with an Overview of Hypertension (Roland Schmieder) and an Understanding of Measurement Modalities for Blood Pressure (Guido Grassi), an overview of the the Guidelines (Orfeas Liangos), and an overview of What You Need to Do to Test for Secondary Hypertension (Stefan Bertog), Optimize Medical Management (Orfeas Liangos) and Patient Selection to Implement a Renal Denervation Practice (Oliver Vonend, Thomas Schmitz).

After these presentations, it was time to take a step back to discuss the anatomy and physiology by such renowned scholars as Dagmara Hering, Ariel Sverdlik, and Paul Sobotka. We took a look at: Renal Denervation and Pulse Wave Velocity (Kai Mortenson), and Clinical Endpoints (Robert Melder).

A special “first-in-man” experience of a new device with impressive results was presented by Dr. van der Heijden, proving how there was still much hope and promise for the future.

After lunch, it was time to take a “deep dive” into the different companies and their current updates: Simplicity (Marc Sapoval and Justin Davies), Vessix (Gerard Goh), Enlighthn (Vasiolos Papademetriou), Covidien (Stefan Bertog), and Paradise Recor Medical (Joost Daemen).

Dr. Darrel Francis came and stole the show with simple explanations as to why Simplicity 3 turned out the way it did. He explained in-depth why the press releases were ill-worded and badly interpreted. This was “irrational exuberance” followed by “unjustified despondency,” and stated that now is the time for an exciting future-based on science. He said all this was due to: “Big Day” bias, “Check Once More” bias, and “I’ll Take Them Now” bias that had harmed this trial. There were solutions to all of these issues that future trials would have to pay attention to if they were to have any hope of success.

The other effects of renal denervation were the focus of the next presentations. These

***“Recent trial data has called the future of renal denervation into question, and this was discussed extensively.”***

presentations included: This included: “Effects on Arrhythmias” (Jennifer Franke), “Insulin Resistance” (Dagmara Hering), “Chronic Kidney Disease” (Roland Schmieder), “Sleep Apnea” (Aleksander Prejbisz), “Exercise Tolerance” (Mathias-Christoph Brandt), “Renovascular Hypertension” (Yvonne Bausback), and “Psychometric Aspects/Quality of Life” (Dagmara Hering).

Once we reviewed all of the above, it was time for a step-by-step approach to the different devices. This started with a lecture on Angiography (Mark Wholey), followed by Instructions for Symplicity Flex and Covidien One Shot (Martin Bergmann), Vessix (Gerard Goh), Enlighthn (Vasiolos Papademetriou), and Recor Medical (Joost Daemen). This was followed by a talk on Invasive Imaging Results after the Four Different Devices (Joost Daemen) and a talk on Optimal Follow-up (Peter de Leeuw).

## Saturday, February 8, 2014

Saturday started with an exciting mix of talks and cases together.

The talks went beyond the earlier presentations with a discussion of the new fields of baroreceptor stimulation, pulmonary artery denervation, new techniques for renal denervation, and more.

This started with an overview of the exciting new field of baroreceptor activation and other new systemic interventions. “What is Baroreceptor Activation” was covered by Joern Schmitt, followed by Barostim in “Renal Denervation Nonresponders” (Hannes Reuter), “Chronic Kidney Disease Patients” (Michael Koziolok), and “Heart Failure” (Rolf Wachter).

The kidney nerves are not the only nerves that play a major role in hypertension. Paul Sobotka discussed “Carotid Body Ablation,” followed by the “Promise and Current Status of Pulmonary Artery Denervation for Pulmonary Hypertension” (Alexander Rothman and Yao-Jun Zhang).

The subject of new techniques for renal denervation was broken into two parts because

of the wide plethora of devices available. Overview presentations included: “Advantages and Disadvantages of Radiofrequency” (Justin Davies), “Ultrasound” (Martin Bergmann), “Histopathology after Ultrasound” (Joost Daemen). Other techniques included: “Cardiosonic” (Michael Jonas), “Instant Renal Denervation with TIVUS” (Michael Jonas), “Terumo Iberis Renal Denervation” (Benjamin Honton), “Cordis Thermocool” (Martin Bergmann), “Bullfrog Catheter and Radiation Therapy” (Jan Kulow), “Medtronic Spyral” (Erwin Blessing), and “Local Anesthesia” (Simon Lam).

The second part included: “Externally Focused Ultrasound” (Omar Dawood), “Denervx Cooled Microwave Technology” (Stefan Bertog), “Perivascular Chemical Renal Denervation” (Tim Fischell), “Northwind Medical” (Mark Wholey), “Chemical Renal Denervation by Vicristine” (Christodoulos Stefanadis) and “VERVE Medical with an Invasive Non-vascular Approach” (Richard Heuser).

Other ‘systemic interventions’ included: “Vagus Nerve Stimulation on Arterial Hypertension” (Sekib Sokolovic), “ROX Medical AV Anastomosis Therapy” (Paul Sobotka) and “The Future of Neurohumoral Modulation.”

Cases, performed by Drs. Horst Sievert, Laura Vaskelyte, and Sameer Gafoor included: live detailed step-by-step discussion of the Recor Medical Paradise, Medtronic Symplicity, Boston Vessix, St. Jude Enlighthn, and Terumo Iberis devices. Live invasive imaging was performed to help with decision management and discussion among the participants was extensive.

This amazing *TRENDS* conference happens yearly in Frankfurt and will happen again in February 2015. We look forward to more exciting developments and discussion and invite you to be part of this conference as we discuss and grow in this exciting field.

For more information visit: [www.csi-congress.org/trend-workshop.php?go=0](http://www.csi-congress.org/trend-workshop.php?go=0). Selected lectures will be found online as well.

**CCT**

Sameer Gafoor, MD  
Cardiovascular Center Frankfurt  
Frankfurt, Germany

[s.gafoor@cvcf Frankfurt.de](mailto:s.gafoor@cvcf Frankfurt.de)



# Medical News, Products & Information

## Winthrop University Hospital Selects Digisonics Cardiovascular Information System as Enterprise Solution

Winthrop University Hospital in Mineola, NY has selected the Digisonics Cardiovascular Information System (CVIS) as their enterprise solution for echo (adult, pediatric and fetal), cath (adult and pediatric), nuclear, and cardiac MR. The Digisonics CVIS will be implemented at three facilities across their enterprise: Winthrop University Hospital, Winthrop Cardiology Associates and Island Cardiac Specialists. The Digisonics cardiovascular information system provides Winthrop's clinicians with a comprehensive clinical database, high-powered PACS and structured cardiology reporting.

Winthrop University Hospital's cardiologists will also utilize Digisonics' newest application, WebView 4.0, which will provide them with the convenience of anywhere, anytime access to their patient images and reports. This application provides users with zero footprint review from any HTML5 capable browser or device. There is no need for ActiveX or other plug-ins and no patient health information is retained on the viewing device.

Seamless integration of the Digisonics Cardiovascular Information System with Philips QLAB, TomTec, Medis and INVIA will provide Winthrop University Hospital with advanced image quantitation and analysis for their cardiovascular studies. A single click launches the 3rd-party analysis software directly from the Digisonics cardiovascular reporting workstation and the post-processed images are saved back into the Digisonics CVIS for review. Winthrop's cardiovascular information system will be enhanced with the Digisonics Search Package, a comprehensive, user-configurable search engine. This powerful tool enables users to quickly set up search criteria to extract clinical information from their cardiovascular information system database for research purposes, compile statistics required for accreditation and generate management reports to target areas for improvement.

HL7 interfaces between Winthrop's Siemens Envision and GE Centricity systems and the Digisonics cardiovascular information system will create a fully electronic and efficient workflow. Winthrop University Hospital will implement Digisonics DigiConnect, a new application enabling the facility to launch any 3rd party systems (such as EMR/HIS, PACS or ECG management software) directly from the Digisonics cardiovascular information system workstation. Users benefit from the convenience of a single sign-on as user credentials and patient information are passed directly to the 3rd party systems. The result is a seamless cardiovascular reporting workflow with improved efficiency, accuracy and turnaround times for all facilities within Winthrop University Hospital's enterprise.

For further information, visit: [www.digisonics.com](http://www.digisonics.com).

## Adult Congenital Heart Association Names New Executive Director - Seasoned Leader Brings More Than Three Decades of Experience to New Role

The Adult Congenital Heart Association (ACHA) - [www.achaheart.org](http://www.achaheart.org), a national nonprofit organization dedicated to improving and extending the lives of adults with congenital heart defects (CHD), announced the

appointment of Glenn R. Tringali as its new National Executive Director.

Tringali comes to ACHA with more than three decades of extensive experience working with small, mid-sized and large national and international healthcare organizations. He succeeds Gail Ober, who led ACHA as Interim Executive Director since October 2013.

"I am honored to join the Adult Congenital Heart Association as National Executive Director," said Tringali. "With more than a million adults in the United States living with congenital heart defects, there is a clear and compelling need for greater awareness and support of ACHA's critically important mission and programs. I am truly excited to have this opportunity to work in partnered leadership with our Board of Directors, Medical Advisory Board, staff and volunteers in making a meaningful difference in the lives of those affected by CHD."

ACHA Board Chair John Fernie commented, "The majority of those born with congenital heart defects are unaware of the need for continued care. An important aspect of our mission is to reach out, support and educate all of those with CHD. Glenn's demonstrated success in expanding outreach, member support and funding brings strong leadership to these critical efforts."

Tringali began his nonprofit career with the March of Dimes, where he served in Executive and Regional Director positions over the course of 10 years. He then honed his skills in development, volunteer coordination and fundraising events in various positions at organizations such as The National Conference for Community and Justice, the Cancer Research Institute and Juvenile Diabetes International. He also served as a Senior Manager Nonprofit Consultant for KPMG Peat Marwick.

He later became the Chief Executive Officer of the National Alliance for Autism Research (NAAR), during which time he managed the expansion of research, development, public relations and government advocacy programs. Under his leadership, NAAR grew from a \$4.2 million organization with a few thousand supporters to a \$13.1 million organization with tens of thousands of supporters throughout the United States, Canada and the United Kingdom. Tringali also grew NAAR's research program commitments by \$6 million. This exemplary success led to the successful merger of NAAR with Autism Speaks, where he assumed the role of Executive Vice President.

Since 2010, Tringali served as the CEO of the Cerebral Palsy International Research Foundation, working to advance the organization's research programs, community awareness and operational funding needs.

"Glenn has a fantastic track record of proven success and his breadth of experience is remarkable. This is an exciting new chapter for ACHA," added Dr. Curt Daniels, Chair of ACHA's Medical Advisory Board and Professor of Internal Medicine and Pediatrics in the Division of Cardiology at The Ohio State University and Division of Pediatric Cardiology at Nationwide Children's Hospital.

Tringali received his undergraduate degree from Rutgers University. He currently resides in central New Jersey.



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## Study Evaluates Role of Infliximab in Treating Kawasaki Disease Antibody Treatment Helps Children with Dangerous Heart Disorder

Newswise — Kawasaki Disease (KD) is a severe childhood disease that many parents, even some doctors, mistake for an inconsequential viral infection. If not diagnosed or treated in time, it can lead to irreversible heart damage.

Signs of KD include prolonged fever associated with rash, red eyes, mouth, lips and tongue, and swollen hands and feet with peeling skin. The disease causes damage to the coronary arteries in a quarter of untreated children and may lead to serious heart problems in early adulthood. There is no diagnostic test for Kawasaki disease, and current treatment fails to prevent coronary artery damage in at least one in 10 to 20 children and death in one in 1,000 children.

Between 10 and 20 percent of patients with KD experience fever relapse following the standard therapy with a single infusion of intravenous immunoglobulin (IVIG) and aspirin. It is known that IVIG resistance increases the risk of heart damage, most commonly a ballooning of the coronary arteries called aneurysms. These children require additional therapy to interrupt the inflammatory process that can lead to damage of the coronary arteries.

A study led by physicians at the University of California, San Diego School of Medicine and Rady Children's Hospital-San Diego looked at intensification of initial therapy for all children with KD in order to prevent IVIG-resistance and associated coronary artery abnormalities by assessing the addition of the medication infliximab to current standard therapy. The results of their study will be published in the February 24, 2014 online issue of the medical journal *Lancet*.

Tumor necrosis factor  $\alpha$  (TNF $\alpha$ ) is a molecule made by the body that plays a role in the development of inflammation in KD; therefore, treatment with a TNF $\alpha$  antagonist is a logical therapeutic intervention, according to the researchers. Early experience with infliximab – a monoclonal antibody that binds TNF $\alpha$  – showed promising results. A Phase 1 trial in children with KD and persistent fever following standard therapy found no infusion reactions or serious adverse events, and subsequent studies suggested that infliximab led to faster resolution of fever and fewer days of hospitalization than a second IVIG infusion.

The UC San Diego researchers conducted a trial of 196 subjects at two centers – Rady Children's Hospital-San Diego, a research affiliate of UC San Diego School of Medicine, and Nationwide Children's Hospital in Columbus, Ohio – to assess whether infliximab could reduce IVIG treatment resistance.

"While the addition of infliximab to primary treatment in acute KD did not reduce treatment resistance, it was safe and well-tolerated, achieved a greater reduction in the size of the left coronary artery, and reduced the number of days of fever and laboratory markers of inflammation," said the study's first author, Adriana H. Tremoulet, MD, of the UC San Diego Department of Pediatrics and the UC San Diego/Rady Children's Hospital-San Diego Kawasaki Disease Research Center. "We conclude that use of infliximab is safe in infants and children and that early treatment could help children with Kawasaki Disease with high levels of inflammation or early signs of coronary artery damage."

## MEDICAL MEETING FOCUS

### PICS ~ AICS 2014

Jun. 7-10, 2014; Marriott Chicago Downtown, Chicago, IL USA

[www.picsymposium.com](http://www.picsymposium.com)

PICS~AICS includes a one day program on June 7<sup>th</sup> - *Cardiac Imaging for Structural Heart Disease* with Course Directors: Roberto M. Lang, MD, FASE, FACC, FESC, FAHA, FRCP; Ziyad M Hijazi, MD, MPH, FACC, FSCAI along with an International faculty.

**PICS Overview:** Taped case sessions, along with the hands-on demonstrations. The meeting continues to support open dialogue as well as visual learning, along with updates from all the relevant trials and societies, with opportunities for networking and sharing global clinical and research experience. The atmosphere is professional, but relaxed. "Tips and Tricks" session with hands-on demonstrations from international experts on how to work with wires, stents, devices and sheaths as well as valves. This will be followed by an interactive taped case session with four cases for discussion. The oral abstract sessions are not competing with one another so that the true scientific endeavors of our colleagues are given the platform they deserve.

**PICS Course Directors:** Ziyad M. Hijazi, MD, MPH; John P. Cheatham, MD; Carlos Pedra, MD; Thomas K. Jones, MD

**Course Co-Directors:** Damien Kenny, MD; Giacomo Pongiglione, MD; Clifford J. Kavinsky, MD, PhD; Ralf Holzer, MD; John D. Carroll, MD

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**Cardiac Imaging Faculty:** Steven Goldstein, MD; Rebecca Hahn, MD; Bijoy Khandheri, MD; Itzhak Kronzon, MD; Amit Patel, MD; Ernesto Salcedo, MD; Vinod Thourani, MD; John Carroll, MD; John Hibblen, MD

**Some of the Topics Covered Include:** Interventional Platform – Tips, Tricks and Application to CHD; Interventional Radiology; Interventional Neurosurgery; Use of CTO Wires in CHD; Hands-on-Demonstrations: Sheaths, Transseptal, Devices, Stents, Valves; My Nightmare Case in the Cath Lab; Complications of Congenital Cardiac Catheterization, and more....

**Breakout Sessions Include:** Complex and New Structural Interventions; Nursing and Associated Professionals; Spanish Session; The Left Atrial Appendage State-of-the-Art; PICES; (Young Interventionalist Group); Mitral Valve Interventions; TAVR

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Additional contributors to the study include: principal investigator Jane C. Burns, MD, Susan Jimenez-Fernandez, MD, John T. Kanegaye, MD, and Beth Printz, MD, of UC San Diego Department of Pediatrics and Rady Children's Hospital-San Diego; Sonia Jain, PhD, and Xiaoying Sun, MS, UCSD Department of Family and Preventive Medicine; Joan M. Pancheri, RN, of y Children's Hospital-San Diego; and Preeti Jaggi, MD, John P. Kovalchin, MD, and Octavio Ramilo, MD of Nationwide Children's Hospital and The Ohio State University, Department of Pediatrics.

Funding for the study was provided by the U.S. Food and Drug Administration, Robert Wood Johnson Foundation, and Janssen Biotech, Inc.

### Clinical Opinion Published on Use of Maternal Oxygen During Labor

When a fetal heartbeat pattern becomes irregular during labor, many practitioners give oxygen to the mother. But questions remain whether this oxygen supplementation benefits the fetus or may actually be potentially harmful.

A clinical opinion written by third year resident Maureen Hamel, MD, along with maternal-fetal medicine specialists Brenna Anderson, MD and Dwight Rouse, MD, of the Department of Obstetrics and Gynecology at Women & Infants Hospital of Rhode Island and The Warren Alpert Medical School of Brown University, has been published in the January 10, 2014 online edition of the *American Journal of Obstetrics & Gynecology*.

The manuscript, entitled "Oxygen for intrauterine resuscitation: Of unproved benefit and potentially harmful," aimed to make recommendations about the safety of the use of maternal oxygen supplementation in laboring women.

According to lead author Dr. Hamel, "Maternal oxygen is often given to laboring women to improve fetal metabolic status or in an attempt to alleviate non-reassuring fetal heart rate patterns. However, there are only two randomized trials investigating the use of maternal oxygen supplementation in laboring women. These studies did not find that supplementation is likely to benefit the fetus and may even be harmful."

Based on their research, the team concludes that until it is studied properly in a randomized clinical trial, maternal oxygen supplementation in labor should be reserved for maternal hypoxia (lack of oxygen) and should not be considered an indicated intervention for non-reassuring fetal status.

### Kawasaki Disease and Pregnant Women - UC San Diego Researchers Say Risks Are Manageable, Provided Doctors Recognize Them

Newswise — In the first study of its type, researchers at the University of California, San Diego School of Medicine have looked at the health threat to pregnant women with a history of Kawasaki disease (KD), concluding that the risks are low with informed management and care.

The findings are published in the March 6, 2014 online edition of the *British Journal of Obstetrics and Gynaecology*.

KD is a childhood condition affecting the coronary arteries. It is the most common cause of acquired heart disease in children. First recognized in Japan following World War II, KD diagnoses are rising among children in Asia, the United States and Western Europe. Predictive models estimate that by 2020 one in every 1,600 American adults will be affected by KD.

"A growing number of women with a history of KD are reaching child-bearing age, but there is little information available to guide their obstetrical care," said study author Jane C. Burns, MD, Professor and Director of the Kawasaki Disease Research Center at UC San Diego and Rady Children's Hospital-San Diego. "By and large, KD is virtually unknown among working obstetricians."

KD is currently diagnosed by a constellation of clinical signs, with supporting lab tests that indicate high levels of inflammation. These signs include abrupt onset of high fever, accompanied by four of five criteria, among them: widespread rash, cracked and fissured lips, "strawberry tongue," bloodshot eyes, lymph node enlargement and red, swollen hands and feet.

Without treatment, 25% of children with KD develop coronary artery aneurysms – balloon-like bulges of heart vessels – that may eventually result in heart attacks, congestive heart failure or sudden death. The condition can be treated with a high-dose of intravenous immunoglobulin and aspirin, reducing the risk of aneurysms to 5%. The long-term risk for adults with a history of KD in childhood is not known.

Senior study author John Gordon, MD, and colleagues conducted the first KD study of non-Japanese patients, and the first to explore the health risks to women with a history of KD and their offspring. They found that the health risks for mothers with no KD-related coronary artery damage were similar to the general population. For women with aneurysms, the risks were low with appropriate management and care.

"The main message is positive," said Burns. "Women who have had KD can successfully deliver to term without complications. C-sections are not necessarily indicated if they have aneurysms, they can labor normally, if their overall cardiovascular status is OK."

There is a genetic component to KD. The study found that two of the 21 children born to the 10 women with a history of KD also developed the disease. "There is clearly an increased risk in offspring," said Burns, "but the (study) numbers are small so we cannot really calculate a risk until there is a larger population of KD adults who have had children."

Co-authors include: C.T. Gordon, S. Jimenez-Fernandez and C. Shimizu, Department of Pediatrics, UCSD; L.B. Daniels and A.M. Kahn, Department of Medicine, UCSD; M. Tarsa, Department of Maternal and Fetal Medicine, UCSD; T. Matsubara, Juntendo University Urayasu Hospital; and John B. Gordon, San Diego Cardiac Center.

Funding for this research came, in part, from grants from the National Heart, Lung, Blood Institute of the National Institutes of Health (grant HL U54 HL108460), the American Heart Association and the Gordon and Marilyn Macklin Foundation.

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## First Human Totally Endoscopic Aortic Valve Replacements (TEAVR) Reported

Newswise — Surgeons in France have successfully replaced the aortic valve in two patients without opening the chest during surgery. The procedure, using totally endoscopic aortic valve replacement (TEAVR), shows potential for improving quality of life of heart patients by offering significantly reduced chest trauma. It is described in *The Journal of Thoracic and Cardiovascular Surgery*, an official publication of the American Association for Thoracic Surgery.

Endoscopic surgery is already used by cardiovascular surgeons for procedures such as atrial septal defect repair and coronary artery bypass grafting. This leads to faster recovery time and less pain, which improves patients' quality of life.

TEAVR had not been feasible previously because of the currently available designs of stented tissue valves. The recent advent of sutureless bioprostheses mounted on a compressible self-expanding nitinol (nickel titanium) stent, was one of the key factors enabling the surgical team to perform this procedure. Implantation required less than 45 minutes in either patient. Sutureless substitutes are not yet available for the other cardiac valves, like the mitral valve.

"In our institution, we began by adopting the mini-sternotomy technique, involving a small incision through the sternum, as routine. We then transitioned to the right mini-thoracotomy approach, involving a small incision through the thorax, first under direct view, then with an endoscopic camera. Finally we adopted a totally endoscopic technique," explains lead author Marco Vola, MD, PhD, of the Department of Cardiovascular Surgery, Centre Hospitalier Universitaire de Saint-Etienne, France.

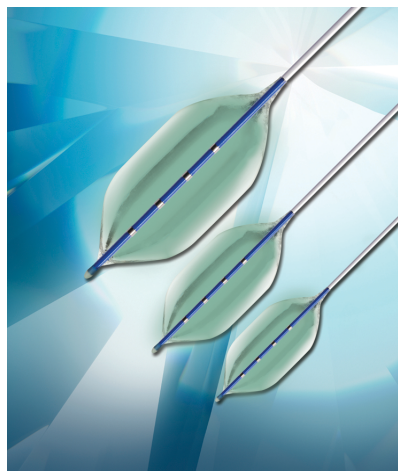
"It is important to note that when performing TEAVR, a quick and safe conversion to mini-thoracotomy under direct view can be made if circumstances demand. This would still offer significantly reduced chest trauma," he adds.

In other fields, totally endoscopic surgery involved longer clamping and cardiopulmonary bypass (CPB) times during the learning curve. The investigators believe that clamping and CPB times were acceptable and that the learning curve could be shorter than reported for totally endoscopic coronary artery bypass grafting.

Enhancements such as endoscopic sizers, dedicated instruments for decalcification, and second-generation sutureless bioprostheses to simplify implantation, could improve the procedure further, Dr. Vola and his colleagues comment. Last but not least, surgical robots may offer additional benefits.

"These first procedures show that totally endoscopic sutureless aortic valve replacement is technically feasible. Further clinical experience and technical development are necessary to shorten operation times and to assess further the potential postoperative benefits of TEAVR," concludes Vola.

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## GW Researcher Finds Gene Therapy a Promising Tool for Cardiac Regeneration

After a heart attack, there is often permanent damage to a portion of the heart. This happens, in part, because cardiac muscle cells are terminally differentiated and cannot proliferate after blood flow is blocked off to the heart. This partial healing can be attributed to heart disease being one of the leading causes of death. What if the cells could be stimulated to divide and the heart could be induced to repair itself? This was the question posed by George Washington University (GW) researcher Scott Shapiro, MD, PhD, and his co-authors, who found that cardiac regeneration may be a possibility with gene therapy.

The research, published in the February 19<sup>th</sup> edition of *Science Translational Medicine*, found that gene therapy can elicit a regenerative response in pig hearts. Shapiro and his research team first looked to small animals such as the zebrafish, which are able to regenerate heart tissue after a heart attack. This animal has a key protein at play, Cyclin A2 (Ccna2).

"After seeing the effects of Ccna2 in small animals, we began looking at the effects of the gene in larger animals, such as pigs," said Shapiro, assistant professor of medicine at the GW School of Medicine and Health Sciences. "We delivered Ccna2 directly into the heart and found that pigs



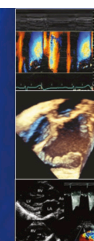
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not only had improved cardiac function, but also found evidence of cellular regeneration."

Ccna2 is a prenatal gene normally turned off in humans after birth. Shapiro believes using gene therapy as a tool for cardiac regeneration, optimized for humans, could lead to a viable treatment option for patients who suffer from myocardial infarction, or heart attack.

The study, titled "Cyclin A2 Induces Cardiac Regeneration After Myocardial Infarction Through Cytokinesis of Adult Cardiomyocytes," is available at <http://stm.sciencemag.org/content/6/224/224ra27.short>.

Additional authors of the study include researchers from the Cardiovascular Institute at the Mount Sinai School of Medicine, the Centro Nacional de Investigaciones Cardiovasculares at the Hospital Universitario La Paz, and the Division of Cardiology at the Albert Einstein College of Medicine.

### **The Children's Cardiomyopathy Foundation (CCF) Announces the Availability of 1-Year Research Grants for Studies Focused on Pediatric Cardiomyopathy**

The purpose of CCF's Research Grant Program is to advance knowledge of the basic mechanism of the disease and to develop more accurate diagnostic methods and improved therapies for children affected by cardiomyopathy. Principal investigators must hold an MD, PhD or equivalent degree and reside in the U.S. or Canada. Funding is available in the range of US\$25,000 to US\$50,000 for one year of total direct costs. CCF requires a Letter of Intent in advance of the grant application. The 2014 deadline for Letters of Intent is June 13<sup>th</sup> by 5:00 p.m. EST. Only investigators who have submitted a Letter of Intent and have been invited to submit a formal grant application will be considered for CCF funding.

The submission deadline for full grant applications is September 5<sup>th</sup>, 2014 by 5:00 p.m. EST with final award decisions to be made in January 2015.

Visit [www.childrenscardiomyopathy.org](http://www.childrenscardiomyopathy.org) (click on Research/Grants & Awards) for application guidelines and to view past grant awards.

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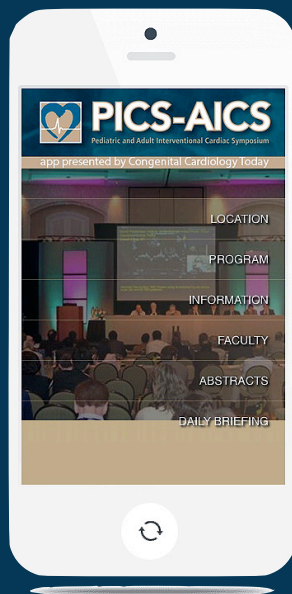
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### **Mailing Address:**

PO Box 444, Manzanita, OR 97130 USA  
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16 Cove Rd, Ste. 200, Westerly, RI 02891 USA

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