

# CONGENITAL CARDIOLOGY TODAY

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

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### Cardiology 2016

Feb. 24-28, 2016; Orlando, FL USA  
[www.chop.edu/events/cardiology-2016](http://www.chop.edu/events/cardiology-2016)

### PICS-CSI Asia 2016 - Catheter Interventions in Congenital, Structural and Valvular Heart Disease

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### SCAI 2016

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## Persistent Unligated Vertical Vein Following Repair of Total Anomalous Pulmonary Venous Return: An Unusually Late Presentation

Brett Larsen, BS; Steven Pophal, MD; Randy Richardson, MD

### Introduction

Total Anomalous Pulmonary Venous Return (TAPVR) is a congenital cyanotic abnormality whereby all pulmonary veins fail to drain into the left atrium, but rather into the right atrium or its venous tributaries. Thus, oxygenated blood is re-circulated through the lungs instead of arterial systemic circulation. The additional presence of a right-to-left shunt defect, such as Atrial Septal Defect (ASD) or Patent Ductus Arteriosus (PDA), is necessary to sustain life.

TAPVR is a relatively rare disease with an incidence of approximately 8 out of every 100,000 births. It is the fifth most common Cyanotic Congenital Heart Disease.<sup>1</sup> Severe disease manifests shortly after birth with tachypnea, cyanosis, and heart failure, while mild disease presents prior to one year with failure to thrive, lethargy, and recurrent respiratory infections.<sup>2</sup> Without surgical intervention, a majority of patients will die within one year.<sup>3</sup>

### Case Report

A 16-year-old male, status post median sternotomy with TAPVR repair at 5 Days of Life presented with precordial chest pain and dyspnea, with concern for possible sternal wire complications. Physical exam revealed stable vital signs with an oxygen saturation of 98%,

normal cardiovascular exam, and point tenderness of the sternum. Echocardiogram revealed a prominent ascending vertical pulmonary vein, which had been a remnant of an incomplete surgical repair. This vertical vein had significant cephalad flow into the superior vena cava and ultimate drainage into the right atrium.

This patient had previously been diagnosed at birth with supracardiac TAPVR with a rare right-sided vertical vein. His systemic circulation of oxygenated blood was preserved via a PDA and an ASD. During his initial surgical repair, all but a single right-sided vertical vein was reconnected to the left atrium. The remaining vein was unable to be ligated due to unusual positioning. A post-op tranesophageal echo showed no evidence of the decompressing vein or pulmonary vein stenosis; thus, the remaining vein was left alone. The patient's post-op course was uncomplicated, and he remained asymptomatic until presentation at 16 years old.

After his presentation, a cardiac MRI and CT chest angiogram with 3D renderings were obtained for further evaluation. Imaging revealed a persistent 2.2 cm patent anomalous vertical vein draining the right-upper and right-middle lobes of the lung, with connections to the superior vena cava adjacent to the junction of the brachiocephalic vein and inferiorly with the left atrium (Figures 1-3). There was also associated dilatation of the superior vena cava to 4 cm in diameter (Figure 3) with moderate dilatation of the right ventricle and the right atrium (Figure 4).

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On catheter angiography, all of the venous drainage from the vertical vein was directed toward the superior vena cava, consistent with left-to-right shunt. He subsequently underwent placement of an Amplatzer Vascular Plug II device just before the junction of the anomalous vertical vein and the superior vena cava. Thus far, the patient has shown clinical resolution of symptoms.

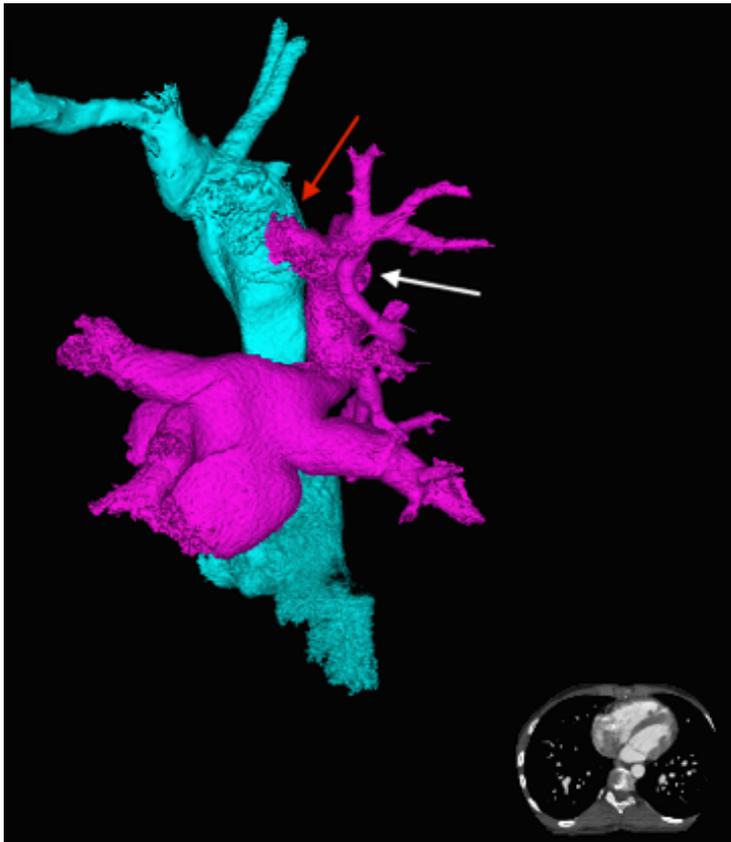


Figure 1. CT 3D reconstruction demonstrating pulmonary veins with left atrium (pink) and systemic veins with right atrium (blue). Also shown are the vertical vein (white arrow) and tributaries with connection (red arrow) of vertical vein and superior vena cava.

## Discussion

The defect causing TAPVR occurs during the 3<sup>rd</sup>-5<sup>th</sup> week of development when the pulmonary system is forming. The embryologic pulmonary venous plexus fails to connect with the left atrium, which instigates retention of connections to either the primitive cardinal and/or umbilicovitelline drainage systems. The former gives rise to connections to the right brachiocephalic vein, superior vena cava, right atrium, or azygous vein, while the latter gives rise to the portal vein, hepatic vein, or inferior vena cava. Accordingly, TAPVR has been classified based on the site at which the anomalous pulmonary veins terminate.<sup>4</sup>

In the most common classification, Type I (44% of cases), the anomalous pulmonary veins conjoin behind the left atrium to form a vertical vein which terminates at the supracardiac level, forming a

## Pediatric Cardiology

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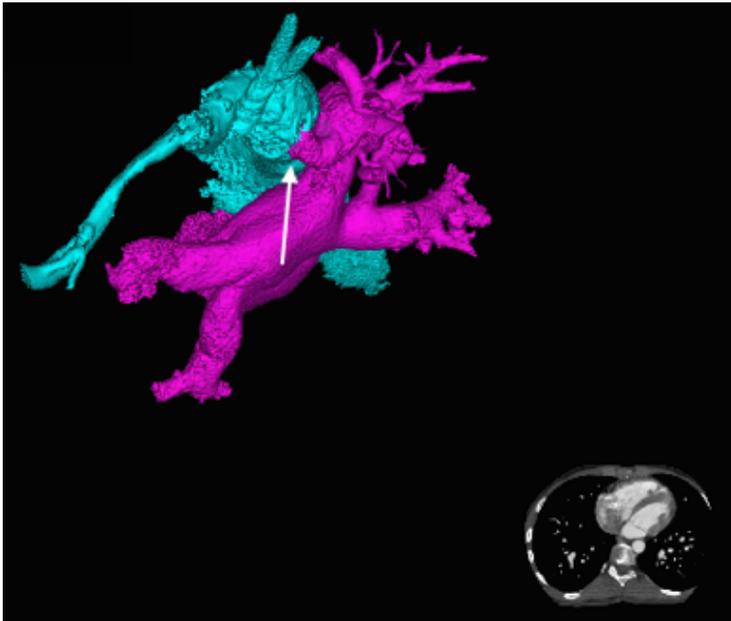


Figure 2. CT 3D reconstruction demonstrating connection of the vertical vein to the superior vena cava (white arrow).

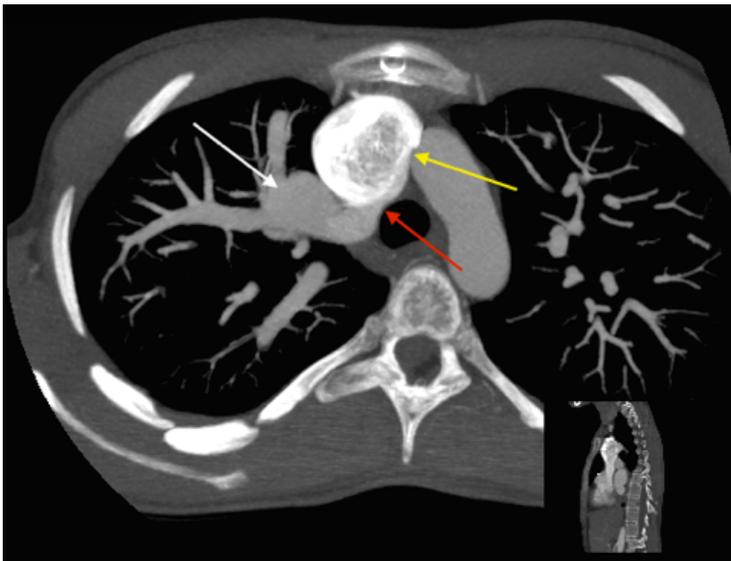


Figure 3. Axial CT demonstrating vertical right-sided vein (white arrow) with anastomosis (red arrow) to the dilated superior vena cava (yellow arrow).

connection with the innominate vein, superior vena cava, or azygous vein. Typically, this vertical vein persists on the left side. In a rare variant, the right cardinal system will persist to form a right vertical vein, which proved to be the case in our patient.<sup>5</sup>

Type II (21% of cases) involves drainage at the cardiac level into the coronary sinus or right atrium. In Type III (26% of cases), the pulmonary veins conjoin into a descending vein that passes below the diaphragm (infracardiac) and terminates at the portal vein,



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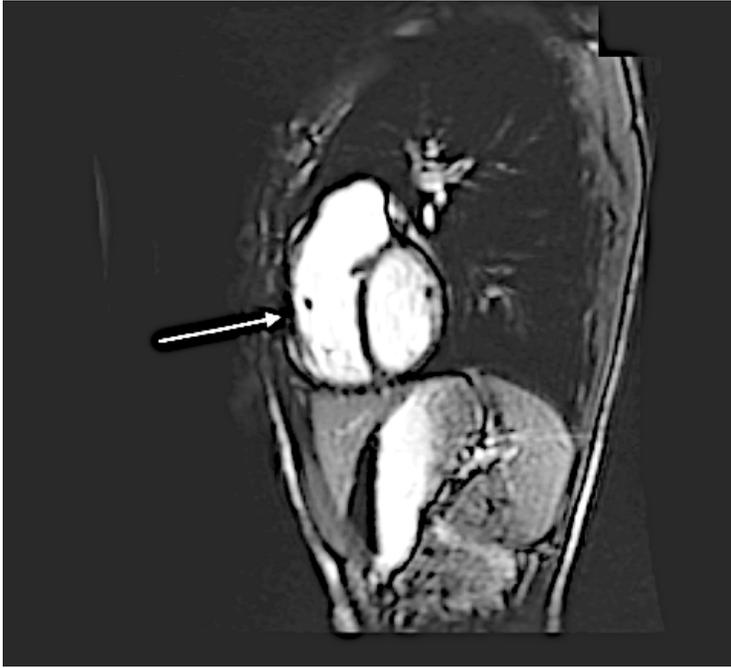


Figure 4. Sagittal MRI demonstrating right ventricular enlargement (white arrow) with mild compression of the left ventricle.

hepatic vein, or inferior vena cava. This type is typically associated with obstruction, causing significant cyanosis at birth. Type IV (9% of cases) involves two or more levels of drainage and is associated with concurrent cardiac abnormalities.<sup>6,7,8</sup>

The natural course of TAPVR yields high mortality rates within the first year of life. Due to the poor prognosis of untreated TAPVR, correction is always recommended regardless of the severity of disease. Treatment is primarily surgical with medical management being utilized for the stabilization in preparation for definitive surgical intervention. Oxygenation, mechanical ventilation, and inotropic support are mainstays of therapy while prostaglandin therapy may be specifically needed to maintain a PDA. In the setting of failed medical stabilization, extracorporeal membrane oxygenation has been utilized to correct metabolic derangements and hemodynamic instability.<sup>9</sup> Previously in the mid 1900's, patient survival fell below 10% following surgical intervention.<sup>8</sup> Survival has dramatically increased over the past four decades with a 1-year survival of 97% in uncomplicated cases and 87% of patients living into the late-teens, with no significant difference between classifications of TAPVR.<sup>10</sup>

In surgical repair of TAPVR, the vertical vein is typically ligated concurrently to avoid persistent left to right shunting.<sup>10</sup> In some specific situations the vertical vein may be left unligated.<sup>11</sup> In these cases, the vertical vein spontaneously closes secondary to preferential flow of pulmonary drainage to the left atrium. There are a small number of cases, such as our patient, where an unligated vertical vein remains patent and eventually leads to significant left to right shunting requiring repair.<sup>11-15</sup> These patients typically developed symptomatic shunting within the first few years after initial repair. The average range of delay between initial TAPVR repair and follow-up presentation with persistent vertical vein was 1-3 years, with the latest case presenting after 5 years.<sup>15</sup> Among the supracardiac TAPVR cases with unligated vertical veins, 50% had patent vertical veins on follow-up with the vast majority requiring further repair. Most underwent surgical ligation while a small minority underwent transcatheter device closure.<sup>15</sup>

## Conclusion

A persistent vertical vein is a rare complication of TAPVR repair with most cases resolving with spontaneous closure. In the minority of

patients where this does not occur, patients typically re-present with symptomatic shunting within 3 years of initial surgery. Our patient proved to follow an unusual course of post-TAPVR repair with persistent right-sided vertical vein. As far as we know this is the first case demonstrating re-presentation as late as 16-years old. His extended asymptomatic post-repair course is unique. It is thought that his pulmonary drainage had preferential flow toward the left atrium with enough shunting toward the superior vena cava, maintaining vertical vein patency, but not causing symptoms. It is unclear what caused his symptomatic reversal of drainage at such a late presentation.

***“Our patient proved to follow an unusual course of post-TAPVR repair with persistent right-sided vertical vein. As far as we know this is the first case demonstrating re-presentation as late as 16-years old.<sup>1</sup> His extended asymptomatic post-repair course is unique. It is thought that his pulmonary drainage had preferential flow toward the left atrium with enough shunting toward the superior vena cava, maintaining vertical vein patency, but not causing symptoms.”***

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# Myocardial Ischemia Caused by an Anomalous Right Coronary Artery: A Case Report

By Tomas Bonilla-Rivera, MD; Jessica Weiss DO; Umaima Fatima, MD

## Abstract

Chest pain and ischemia from an anomalous origin coronary artery is a rare, but described event, especially in the presence of an anomalous Right Coronary Artery (RCA) coursing between the aorta and pulmonary artery. Such a course is termed 'malignant' due to its potential for causing ischemia. We present a case of an anomalous right RCA lying between the aorta and pulmonary artery causing myocardial ischemia.

## Case Report

A 63-year-old male with past medical history of hypertension and dyslipidemia, a family history of coronary artery disease and previous smoking presented with several months of left-sided substernal chest pain and left shoulder pain with moderate activity. Previous cardiac catheterization 12 years ago was negative for Coronary Artery Disease. Transthoracic echocardiogram (TTE) showed new apical hypokinesis and reduced left ventricular ejection fraction of 45%. Electrocardiogram (ECG) showed inferolateral T-wave inversions (Image 1). Stress echocardiogram revealed inferior, basal and septal wall motion abnormalities during exercise, and the patient experienced his typical chest pain symptoms that resolved after recovery. Left heart catheterization revealed patent left main, left anterior descending, ramus intermedius, circumflex and obtuse marginals; however, the RCA was not well visualized. Cardiac CT angiogram was done to further visualize the RCA, and revealed that the RCA had an anomalous origin from the left coronary cusp with a lethal course due to significant compression of the proximal portion between the aorta and the main pulmonary artery (Images 2-4). This finding correlated with the inferior left ventricular wall motion abnormality on stress echocardiogram and the inferolateral ischemic changes on ECG.

The patient refused surgical intervention and chose to be treated with medical therapy for his angina symptoms. He has been medically managed with beta-blockers and avoidance of strenuous physical activity, and has remained asymptomatic for over two years.

## Discussion

Anomalous origin of the RCA from the left coronary sinus has a prevalence of 0.25 – 0.5% in the general population. Most patients

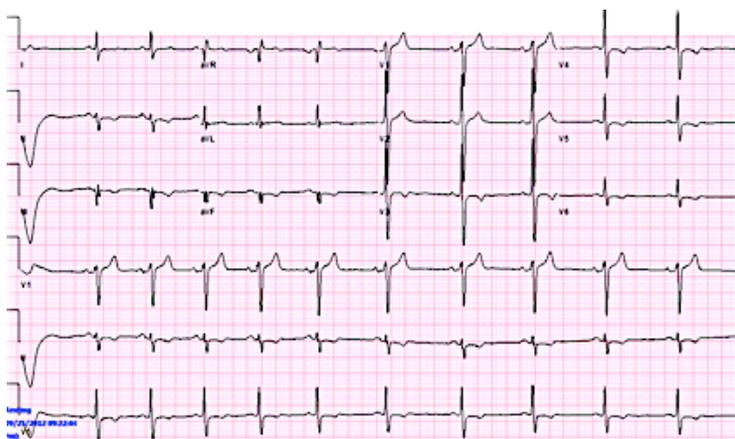
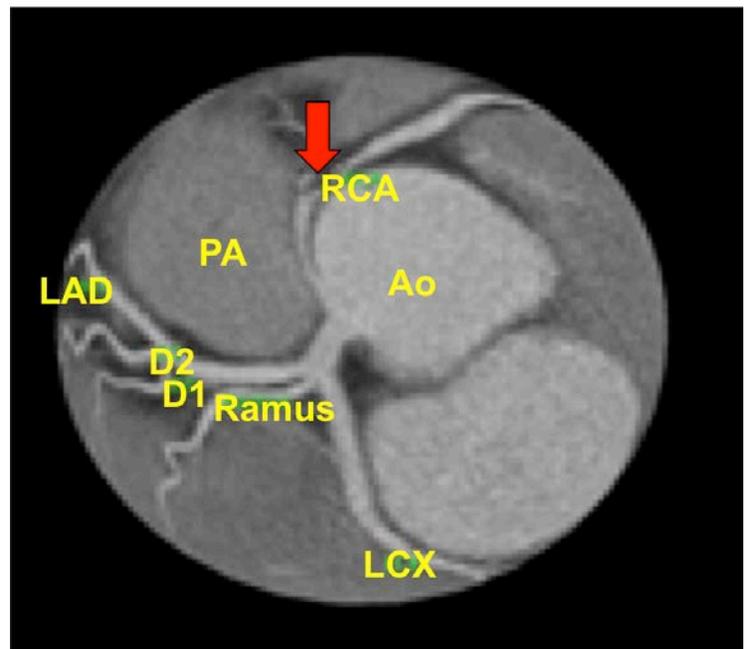
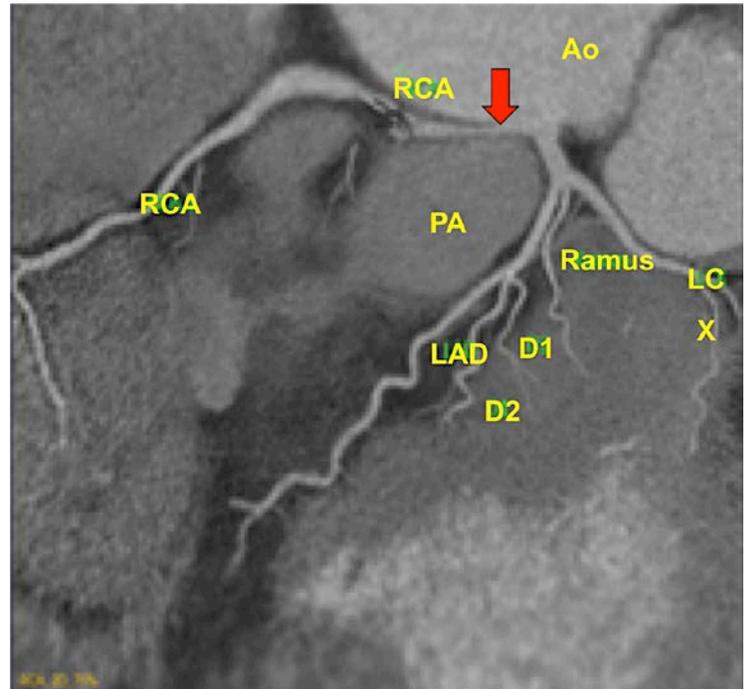


Image 1: Electrocardiogram showing inferolateral T wave inversions in leads II, III, aVF and V3-6.

are asymptomatic throughout their lives or present with sudden death after significant exertion. Cases presenting with chest pain associated with reversible ischemia are exceedingly rare consisting of just a few case reports. Treatment options include: medical management with beta-blockers and avoidance of strenuous activity in asymptomatic patients and surgical bypass grafting as definitive therapy for symptomatic patients with severe stenosis. This case report



Images 2 (top) and 3 (bottom): Coronary Computed Tomography Angiography images showing the anomalous origin of the right coronary artery (RCA) from the left coronary cusp (arrow) adjacent to left main coronary artery (LM). The RCA is compressed between the aorta (Ao) and the pulmonary artery (PA).

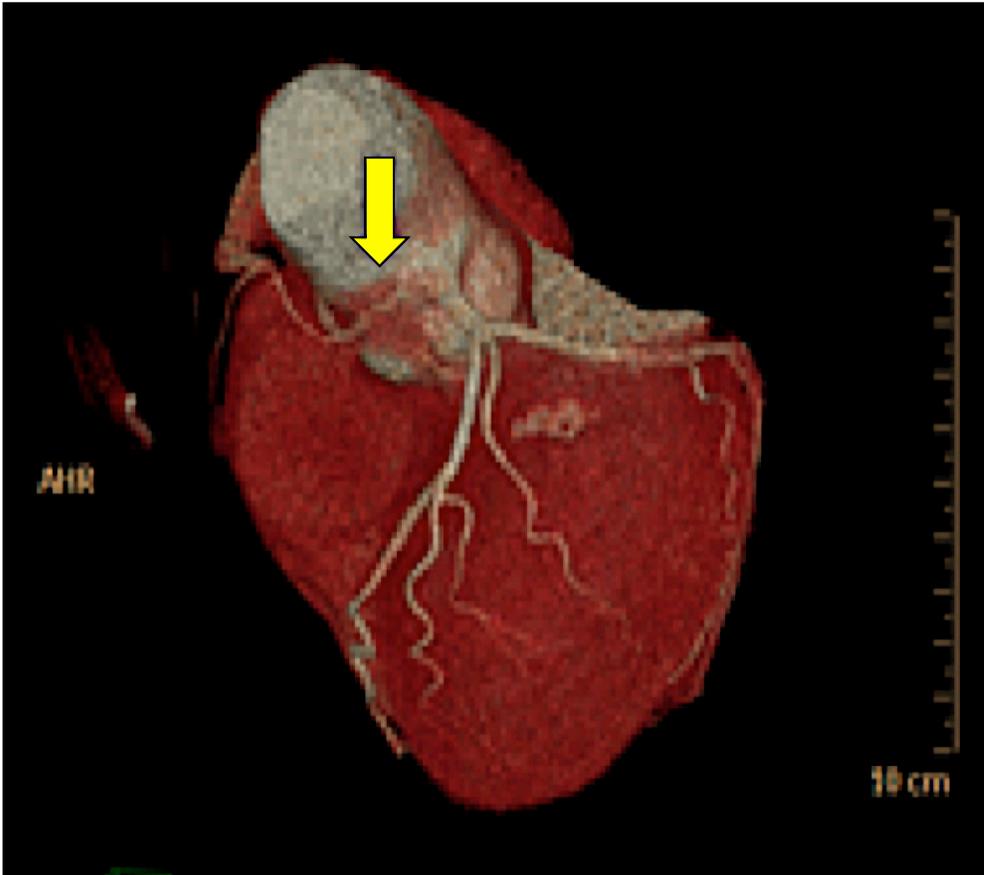


Image 4: Coronary Computed Tomography Angiography 3D reformat image showing the course of THE anomalous right coronary artery RCA (arrow).

is an incredible example of successful medical management of a patient with exertional angina due to severe stenosis of an anomalous RCA compressed between the aorta and pulmonary artery.

There are no disclaimers or financial disclosures to report.

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# The PICES Group: Highlights from Breakout Conferences 2015

By Brent M. Gordon, MD

The Pediatric/Congenital Interventional Cardiology Early-Career Society (PICES) had a very busy 2015. The group held breakout sessions at the SCAI conference in San Diego, the CSI conference in Frankfurt, Germany, and at the PICS/AICS conference in Las Vegas. PICES was established in July 2011, and is currently a sub-committee under the umbrella of the Congenital Heart Disease Council of SCAI. The group was created to support and advance the careers of young interventionalists in the fields of pediatric and adult congenital and structural heart disease. The goals of PICES include: promoting clinical education, fostering multi-center research collaboration, expanding international membership, and creating a professional network of young interventionalists and investigators. The newly-elected PICES executive board is composed of President Nathaniel W. Taggart, MD, (Mayo Clinic, Rochester, Minnesota); Research Chair Jeffrey D. Zampi, MD, (CS Mott Children's Hospital, University of Michigan); Clinical Chair Matthew A. Crystal, MD, (Morgan Stanley Children's Hospital – New York Presbyterian, Columbia University Medical Center, New York); and Secretary Gareth Morgan, MB, BCh, (The Evelina Children's Hospital at Guys and St. Thomas's, London).

The PICES group kicked off the congenital heart meeting at SCAI with a well-attended morning breakout session. Educational lecture series are one of the hallmarks of breakout sessions, and the SCAI breakout session featured talks by Jeffery Darst, MD (Colorado Children's Hospital), and Vivian Dimas, MD, (Children's Hospital Dallas). Dr. Darst shared his center's experience with creating a percutaneous veno-venous ECMO program, and highlighted the necessary steps required to effect change in both a center's culture and treatment strategy. Dr. Dimas updated the group on advances in ventricular assist devices, including the new right ventricular assist device, which may have a larger role in congenital patients. She also touched on the lost art of hemodynamics and the vital role of understanding these processes within the workings of an interventional heart failure team. Bryan H.



Left-to-right: Drs. Brent Gordon, Matthew Crystal, Bryan Goldstein and Gareth Morgan.

Goldstein, MD, (Cincinnati Children's Hospital) provided an update of various PICES-driven multi-center research. Manuscripts in preparation for submission include the periventricular hybrid VSD study, the role of smart technology in the congenital catheterization laboratory, and two manuscripts on stent characteristics and fracture potential of commonly used stents in the treatment of Congenital Heart Disease. The session concluded with Jeffrey Delaney, MD (Omaha Children's Hospital) presenting data from the PICES salary survey, which collected data from almost 50 early-career pediatric interventionalists. Results of the survey were separated by the number of years out of training and disseminated to the group.

The PICES group currently has over 140 U.S. and international members. PICES is very interested in establishing greater membership outside of the United States to facilitate and foster international research collaboration. For the second year in a row, the group was fortunate enough to be awarded a breakout session at the CSI meeting in Frankfurt, Germany. The session was very well-attended, quickly becoming a standing room only affair. Brent M. Gordon, MD, (Loma Linda University Children's Hospital) and Bryan H. Goldstein, MD, gave a synopsis of the group's goals and objectives to the audience, and updated them on ongoing and planned research

***“The group was created to support and advance the careers of young interventionalists in the fields of pediatric and adult congenital and structural heart disease. The goals of PICES include: promoting clinical education, fostering multi-center research collaboration, expanding international membership, and creating a professional network of young interventionalists and investigators.”***

endeavors. The highlight of the session was a talk given by one of the CSI meeting organizers, Shakeel Qureshi (The Evelina Children's Hospital at Guys and St



**Archiving Working Group**  
**International Society for Nomenclature of Paediatric and Congenital Heart Disease**  
[ipccc-awg.net](http://ipccc-awg.net)

Thomas's, London), who spoke about the mentor/mentee relationship. Dr. Qureshi outlined what elements he thought were key in creating a viable and mutually beneficial mentor/mentee pairing. He also stressed that while these relationships can allow one to advance academically, they are also vital when dealing with bad outcomes and complications in our patients. A very emotional and open debate followed that all participants found extremely valuable. The session wrapped up with a case presentation of an embolized muscular VSD device by Sebastian Goreczny, MD (Polish Mother's Memorial Hospital, Lodz, Poland). The group followed the breakout session with a dinner from Cook Medical, and was treated to a talk by Oliver Stumper, MD, (Birmingham Children's Hospital) about complex branch pulmonary artery stenting in infants, which focused on his work with Formula stents.

The PICES group finished off the year with a breakout session at the *PICS~AICS* meeting in Las Vegas, Nevada. Elchanan Bruckheimer, MD, (Schneider Children's Medical Center, Israel) was the keynote speaker at this year's breakout session with his talk entitled, "Bringing Holographic Imaging from the Idea to the Cath Lab." Dr. Bruckheimer discussed his pioneering work with developing and prototyping holographic imaging for real-time usage in the cardiac catheterization laboratory. He also touched on the importance of a team-based approach and involving our interventional radiology, ENT and pulmonology colleagues when treating our more complicated patients. PICES was also fortunate enough to have a case presentation from Michael Seckeler, MD, MSc, (Sarver Heart Center, Tucson, AZ) on fenestration creation in a failing fontan. The case generated lively discussion from the audience and demonstrated numerous teaching points. During the research update, Dr. Goldstein noted that Dr. Seckler's manuscript on the role of smart technology in the congenital cath lab was recently accepted to *Congenital Heart Disease*. Three other manuscripts are to be submitted in the fall. The group finished off with an educational lecture on the US experience with the Gore Cardioform Septal Occluder by Joseph Paolillo, MD, (Sanger Heart and Vascular Institute – Charlotte, North Carolina). Attendees followed up the didactic session with a hands-on workshop where they were allowed to implant the Cardioform Septal Occluder in porcine hearts.

The PICES email listserv is used for clinical discussion, planning research projects, and as a forum for communication among its members and with the PICES Executive Board. The PICES website can be accessed from the SCAI homepage under the "About SCAI" section and "Committee" subsection. The next formal PICES meeting will be in May 2016 at SCAI in Orlando. For further information, or to be added to the PICES listserv, please contact Gareth Morgan at: [drgarethmorgan@gmail.com](mailto:drgarethmorgan@gmail.com).

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Corpus Christi and the Rio Grande Valley offer a relaxed "island style" setting with miles of Gulf beaches and mild weather perfect for outdoor activities. South Texas offers world class hunting, fishing, sailing and wind surfing. Golf, cycling and tennis are enjoyed year round. The cost of living in south Texas is low, and there is no state income tax.

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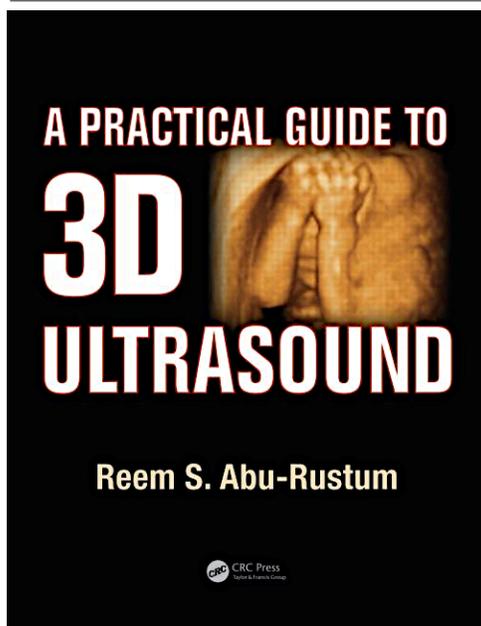
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# Book Review: "A Practical Guide to 3D Ultrasound," by Reem S. Abu-Rustum

Reviewed by Mark S. Sklansky, MD



*A Practical Guide to 3D Ultrasound*  
by Reem S. Abu-Rustum, MD  
December 9, 2014 by CRC Press  
Reference - 173 Pages - 265 Color  
ISBN-13: 978-1482214338  
ISBN-10: 148221433  
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Available in print and for Kindle.

Over the past 25 years, the field and practice of diagnostic obstetric/gynecologic ultrasound has advanced like never before. Much of this progress is due to the development of increasingly powerful computing systems and sophisticated transducer technology. Advances in these areas have resulted in vast improvements in 2D image quality, with exponentially superior depth and resolution capabilities.

At the same time, these dramatic technological advances have also led to the development of profound 3D/4D imaging capabilities. Since the early pioneering work of Dolores Pretorius and Thomas Nelson in the late 1980s and early 1990s, the field of 3D/4D obstetric/gynecologic ultrasound has grown from an intriguing novelty in scattered academic centers, to an important, commercially-viable clinical tool in practices around the world.

While many thousands of papers have been written on 3D/4D obstetric/gynecologic ultrasound, few resources exist for the practitioner interested in a general review of the technique, and in a practical explanation of its clinical application. Dr. Reem S. Abu-Rustum, an internationally recognized authority and wonderfully-gifted teacher in the

field, has recently published an outstanding, clinically-oriented "how-to" textbook to fill this void. *A Practical Guide to 3D Ultrasound* will be of great interest to those practitioners interested in introducing or expanding the use of 3D/4D obstetric/gynecologic ultrasound in their practice or at their institution.

The textbook begins with easy-to-read background chapters on terminology, acquisition, evaluation and display of volume-data. Subsequent chapters discuss the application of specific techniques and software, such as STIC (spatiotemporal image correlation), VCAD (volume computer-aided diagnosis), and VOCAL (virtual organ computer-aided diagnosis), followed by multiple organ-specific chapters detailing the approach and rationale for application of 3D/4D ultrasound to various obstetric/gynecologic and (primarily) fetal areas (heart/spine/brain/face/GI/GU, among others). The text ends with an informative chapter on coding/billing and the role of medical ultrasound practices in providing "keep-sake" fetal images to expectant patients and their families.

This well-written and clinically grounded textbook has innumerable strengths, foremost among them: Dr. Abu-Rustum's years of experience, attention to detail, and candid, practical approach. Beautiful and instructional color images complement the text throughout the book, practically on every page. These images are priceless, and can be reviewed alone along with their legends. Specific descriptions of various published algorithms and techniques can help readers translate what can be found in academic journals to actual clinical practice. Among the greatest assets of the book are the precious "practical pearls" found at the conclusion of each chapter. These pearls alone are worth the price of the text.

The textbook's primary weakness may represent the flip-side of one of its strengths. The inclusion of terminology, approaches and algorithms that are currently vendor-specific will be useful for many readers, but will not be available to all practitioners, and may soon be replaced with newer and more sophisticated algorithms and techniques. This weakness may be most evident in those portions of the text relating to fetal cardiac 3D/4D ultrasound; the clinical application of 3D/4D techniques to the fetal heart remains limited primarily by image quality considerations, but also by somewhat cumbersome vendor-specific algorithms/techniques.

Dr. Abu-Rustum's textbook represents a landmark "user's manual" for the practitioner of 3D/4D obstetric/gynecologic ultrasound, and teaches many valuable lessons beyond specific techniques. Among these lessons, as

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***"Moving forward, practitioners of obstetric/gynecologic ultrasound around the world are sure to see 3D/4D applications continue to have an increasingly important and pervasive role in everyday practice."***

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stated in her "practical pearls" section, is that "the key to a good 3D image is a good 2D image." Dr. Abu-Rustum appropriately emphasizes this caveat throughout the text, as well as the potential for artifact found in all applications of 3D/4D ultrasound. She offers numerous tips/pearls on how to obtain high quality 2D (and thus 3D/4D) images, and how to minimize artifact, including narrowing the field of view and obtaining proper angles of acquisition. But, the book's focus remains squarely on the practice of 3D/4D obstetric ultrasound.

Moving forward, practitioners of obstetric/gynecologic ultrasound around the world are sure to see 3D/4D applications continue to have an increasingly important and pervasive role in everyday practice. Dr. Abu-Rustum's *Practical Guide to 3D Ultrasound* will help us get there.

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# Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

## Digisonics Introduces Seamless Integration Between Cardiovascular Information System Solutions and GE EchoPAC™

Digisonics has introduced seamless integration between its Cardiovascular Information System Solutions (CVIS) and GE Healthcare's EchoPAC for advanced analysis of 3D/4D studies and strain rate. A single click launches the EchoPAC software directly from a study image within the Digisonics CVIS.

The clinical workflow is streamlined with the elimination of having to toggle between two separate workstations and applications. The user can perform the advanced 3D/4D analysis in EchoPAC and store the processed image back into the Digisonics CVIS, facilitating a seamless workflow.

"Digisonics is very pleased to add the GE EchoPAC application to our best-in-class suite of CVIS interfaces," commented Ernest Jackson, Chief Technology Officer. "With EchoPAC, users have the ability to post-process the original RAW data acquired on the ultrasound system, expanding productivity and leveraging the advanced quantitative and multi-dimensional analysis provided."

GE's EchoPAC technology provides image processing, annotation, analysis, measurement, report generation, communication, and storage, and retrieval of ultrasound images that are acquired via the GE Vivid family of ultrasound scanners. The EchoPAC software is an integral component of each Vivid system, providing the post-acquisition image management and reporting functions of the scanner.

Digisonics provides top-rated clinical image management and structured reporting systems for cardiovascular (CVIS), radiology, and obstetrics & gynecology. Digisonics structured reporting solutions combine high performance image review workstations, a powerful PACS image archive, an integrated clinical database, comprehensive analysis capabilities and highly configurable reporting for multiple modalities. Key applications are complemented with interfaces to information systems and 3rd party vendors, providing facilities with a seamless, efficient clinical workflow. For more information: [www.digisonics.com](http://www.digisonics.com).

## Early Surgery for Mitral Regurgitation Before Clinical Triggers Emerge Has Best Outcomes

About 2% of the U.S. population has mitral valve regurgitation, which left untreated, can remain mild or lead to arrhythmia or heart failure. Timing of surgery is a matter of controversy, with guidelines suggesting "watchful waiting" or medical treatment until heart failure or poor function becomes apparent. Now a report in the *Journal of Thoracic and Cardiovascular Surgery*, the official publication of the American Association for Thoracic Surgery (AATS), finds that allowing patients to reach these "surgical triggers" doubles the risk of postoperative mortality and heart failure compared to those who undergo early surgery.

"Our study indicates that early repair should be preferred to rescue surgery in patients with mitral regurgitation (MR)," explained lead author Maurice Enriquez-Sarano, MD, of the Division of Cardiovascular Diseases and Internal Medicine, Mayo Clinic (Rochester, MN). "Guideline triggers for MR surgery based on symptoms and complications are linked to excess postoperative mortality and morbidity versus early surgery. Early surgery in this era of low operative risk and high repair rates provides the best long-term outcomes after MR surgery."

MR is a condition caused by blood leaking backwards through the partially closed mitral valve that connects the two left-sided chambers of the heart. For many people, symptoms may be mild and progress slowly over decades. The sign found on examination is a heart murmur and the symptoms include difficulty breathing and severe fatigue and swollen feet or ankles (heart failure) as well as heart palpitations (atrial fibrillation), but can lead to progressive deterioration of functional capacity associated with excessive pressure in the lungs (pulmonary hypertension).

This study analyzed data from 1,512 patients who were seen at Mayo Clinic between 1990 and 2000 for surgical correction of MR. The average age was 64 years, 89% had mitral prolapse (a condition in which the leaflets and tendons supporting the mitral valve weaken, preventing the valve from closing tightly), and 88% had their valves repaired, rather than replaced.

Patients were divided into three groups according to surgical indication. The first group of 794 patients had Class I triggers, such as heart failure symptoms, an ejection fraction of <60%, or an end-systolic diameter of 40 mm. The second group of 195 patients had Class II triggers, which included complications such as atrial fibrillation or pulmonary hypertension. The third group of 523 patients had severe MR, but no other surgical trigger, and underwent early surgery.

The investigators found that after adjusting for age, the mortality risk was approximately doubled in the Class I group, and increased by 40% in the Class II group compared to those who underwent early surgery. The 15-year survival was 42% in the Class I group, 53% in the Class II group, and 70% in the early surgery group.

Patients may defer surgery because of anxiety about the procedure. Overall the operative mortality was 0.6% which was lower than the 2.5% predicted. The only group with operative mortality was the Class I indication group, which had a rate of 1.1% compared to no deaths in the Class II or early surgery groups. Hospital stay was also longer for those in the Class I (10.6 days) and Class II (10.5 days) groups compared to the early surgery (8.3 days) group.

During follow-up, 302 patients developed postoperative heart failure. After multivariate adjustment, an approximately 2.5 increased risk was found for heart failure in the Class I trigger group and a two-times increased risk was found in the Class II trigger group. Significant differences between groups were found for the combined end-point of death or heart failure: the rates were 37% in the early surgery group versus 65% in the Class I trigger group and 55% in the Class II group. "Early surgery for those with severe MR performed before clinical triggers develop provides the best operative outcome and lowest mortality/congestive heart failure," noted Dr. Enriquez-Sarano.

"Although this very carefully analyzed large series from a single institution is not definitive, the findings are indeed concerning that current clinical practice and guidelines are in fact putting larger numbers of patients at unnecessary risk of death and premature heart failure. If history continues to repeat itself, these data probably will be confirmed by other large centers and will eventually result in adjustment to the guidelines," commented Donald D. Glower, MD, of the Department of Surgery, Duke University Medical Center (Durham, NC) in an accompanying editorial.

Dr. Glower agrees that the early surgery approach should be reserved for higher-volume institutions and patients with a high likelihood of repair. He urges "physicians to be honest enough to look at themselves, and not just at the patients" when making decisions about when, where, or by whom patients with MR should undergo surgery.

## Studies Find That Delayed Umbilical Cord Clamping May Benefit Some High-Risk Newborns

Newswise — Clamping and cutting of the umbilical cord happens within 10 seconds after birth in most cases, in part so members of a medical team can more quickly begin caring for a newborn. But research from Nationwide Children's Hospital shows that waiting 30 to 45 seconds to clamp could have advantages for extremely preterm infants.

The study, published online September 24<sup>th</sup> in the *Journal of Perinatology*, found that the preterm infants with delayed cord clamping had higher blood pressure readings in the first 24 hours of life and needed fewer red blood cell transfusions in their first 28 days than infants whose umbilical cords were immediately clamped. In addition, the short delay made no difference in the safety of the infant immediately after delivery.

The study examined 40 infants who were born at a gestational age between 22 and 27 weeks. A baby is considered to be full term at 39 weeks; 22 weeks is considered the limit of viability. The average birth weight of the babies studied was approximately 1.4 pounds.

"Infants born prior to 28 weeks gestation represent a high-risk subgroup, so efforts to improve outcomes remain critically important," says Carl Backes, MD, a cardiologist and neonatologist at Nationwide Children's, and lead author of the study. "There is increasing evidence that delayed cord clamping may give infants in many categories a better chance."

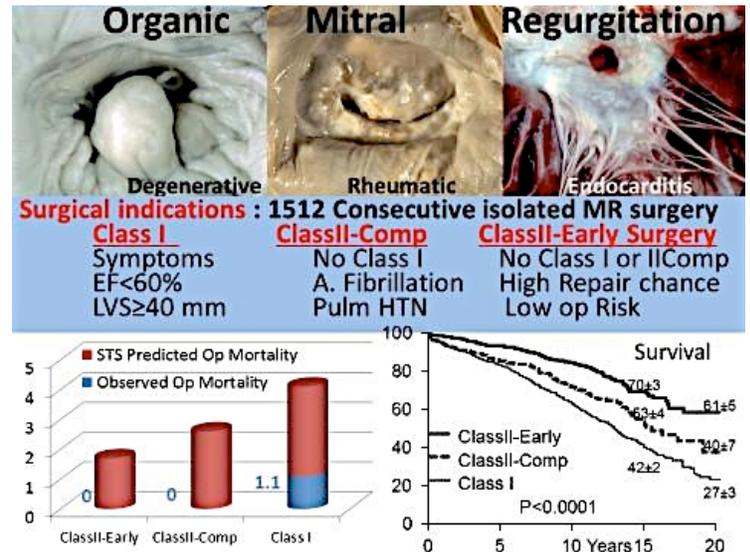
Dr. Backes also led a study, published in July in the *Journal of Perinatology*, which found delayed cord clamping may be beneficial for newborns with Critical Congenital Heart Disease. In that study, infants whose umbilical cords were clamped approximately two minutes after birth needed fewer red blood cell transfusions than infants whose cords were clamped within 10 seconds.

The delay allows for an increased blood volume in the baby, which likely improves pulmonary blood flow and other circulatory measures, stabilizing blood pressure. This may be particularly important for infants with critical congenital heart disease according to Dr. Backes, who is also an assistant professor of Pediatrics at The Ohio State University College of Medicine.

"Further research is needed in both of these infant populations to see whether the short-term benefits translate to reductions in long-term morbidity," Dr. Backes says. "The early results are promising, though."

## Surgeons Refine Procedure for Life-Threatening Congenital Heart Defect

For children born with life-threatening Hypoplastic Left Heart Syndrome (HLHS), reconstructive surgeries can restore blood circulation. While the most common corrective approach is the three-stage Norwood procedure, an alternative strategy, hybrid palliation, allows deferral of the more complex reconstructions to when the child is somewhat older and better able to successfully recover from major surgery. A report in *The Journal of Thoracic and Cardiovascular*



*Journal of Thoracic and Cardiovascular Surgery.*

*Surgery*, the official publication of the American Association for Thoracic Surgery (AATS), evaluates whether an arterial shunt in the hybrid palliation may be a better source for the pulmonary blood supply than the more frequently used venous shunt.

Each year, almost 1000 babies in the United States are born with HLHS, a congenital condition in which the left side of the heart is undeveloped and systemic blood flow is inadequate to sustain life. Without surgical intervention, either reconstruction of structures of the heart and blood vessels or cardiac transplantation, HLHS is fatal. Symptoms of HLHS manifest hours or days after birth, when the ductus arteriosus, a blood vessel connecting the pulmonary artery and the aorta, begins to close. Symptoms include breathing problems, pounding heart, weak pulse, ashen or bluish skin, and heart murmurs. The three-stage Norwood procedure occurs at different times of development, with Norwood Stage I typically performed soon after birth, Norwood II (commonly referred to as a bidirectional Glenn - or cavopulmonary - shunt) between 4 and 6 months of age, and the final surgery, termed a modified Fontan Procedure, between 2 and 5 years of age.

Although Norwood palliation has achieved 30-day survival rates of 90% or more, surgeons strive for better ways to improve outcomes for the highest risk patients. "Hybrid palliation was initially thought to be a therapy that would eventually supplant standard Norwood palliation because of its technical simplicity, its avoidance of cardiopulmonary bypass (open heart surgery) and prolonged perioperative recovery in the neonatal period, and an intuitive notion that it would be associated with improved neurodevelopmental outcomes," commented David M. Overman, MD, Chief of the Division of Cardiovascular Surgery at the Children's Hospitals and Clinics of Minnesota (Minneapolis) in an accompanying editorial.

"In our center, the hybrid procedure is reserved for higher-risk, more complex, and unstable patients in whom a traditional Norwood procedure would carry an unacceptably high risk," explained lead

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investigator Mohamed S. Nassar, PhD, FRCS, of the departments of Paediatric Cardiology and Cardiac Surgery, Evelina London Children's Hospital, Guy's and St. Thomas' NHS Foundation Trust (London).

In the second stage of the hybrid strategy, the aortic arch must be reconstructed and a source of blood supply to the lungs must be established. In a quest to refine the hybrid procedure, surgeons may use two possible sources for the pulmonary blood supply. The usual practice was to create a venous shunt between the pulmonary artery and superior vena cava (a cavopulmonary shunt). However, other investigators have raised concerns about a venous shunt, preferring instead an initial arterial shunt constructed between the first branch of the aortic arch and the right pulmonary artery (modified Blalock-Taussig shunt), followed later with the superior cavopulmonary anastomosis.

"The clinical issue driving this novel strategy is the increasingly well-documented problem of branch pulmonary artery stenosis associated with hybrid palliation of HLHS," noted Dr. Overman. "The authors' premise that use of an arterial shunt at the time of arch reconstruction may result in improved pulmonary artery architecture is reasonable."

Since both the arterial and venous shunts were being performed at the same institution, the researchers had an ideal opportunity to conduct a retrospective analysis of their cases to see whether one procedure offered better outcomes than the other. Dr. Nassar identified 17 HLHS patients who received an arterial shunt, and 26 patients who received a venous shunt. Indeed, the arterial group did show better pulmonary arterial growth than the venous shunt, as indicated by a higher lower lobe index.

There are other pros and cons to each procedure. Surgical times were shorter in the arterial group, but this group also demonstrated a higher need for delayed sternal closure. Mechanical ventilation and intensive care stay were shorter in the venous shunt group. At discharge, there was no difference in the echocardiography findings, but those who received arterial shunts had significantly higher oxygen saturations.

No differences were found in mortality or incidence of complications between the two approaches. Both groups had approximately a 30% rate of MRI-proven brain abnormalities after clinically detected abnormal neurological findings in the immediate postoperative period. "This relatively high incidence is comparable to other reports studying the neurologic development in patients with HLHS," stated Dr. Nassar.

Dr. Overman expressed his concern with the high rate of clinically detectable postoperative neurologic abnormalities and associated MRI findings in both treatment groups. "The central tenet of hybrid strategy is that neurodevelopmental outcomes will be improved by delaying larger reconstructive operations from the neonatal period into infancy."

While Dr. Overman noted that hybrid surgery strategies for HLHS are currently used only for a minority of patients, with the Norwood procedure still being preferred at most institutions, he acknowledged that there is a place for hybrid surgery in a higher-risk subset of



## Pediatric Cardiac Intensivist

The Cardiology Division of the Department of Pediatrics at LSU Health Sciences Center in New Orleans is accepting applications for full-time academic pediatricians or pediatric subspecialists with specialized training and expertise in cardiac critical care. Appointment will be at the academic rank of Assistant Professor, Associate Professor, or Professor (non-tenure, clinical track) and will be determined by the candidate's credentials and experience.

The successful candidate will direct the medical management of cardiac patients requiring intensive care and of patients having undergone surgery for congenital and/or acquired heart disease. The primary site of practice will be Children's Hospital New Orleans. The intensive care team will work in conjunction with pediatric cardiologists, cardiothoracic surgeons and other subspecialists, in the management of patients.

Currently, approximately 250 heart catheterizations (75% interventional) and 350 cardiothoracic surgeries are performed each year at Children's Hospital New Orleans. Children's Hospital is a 247 bed tertiary-care teaching complex located in the heart of New Orleans, which serves the entire State of Louisiana and the central Gulf Coast region. Children's Hospital has a state-of-the-art 20 bed CICU. There is an ACGME-accredited LSUHSC pediatric cardiology fellowship training program. There are excellent opportunities for clinical, translational and basic research, particularly with the LSUHSC Cardiovascular Center of Excellence. The successful candidate is expected to have strong clinical skills, have an interest in research, and participate in teaching of residents and fellows.

Qualified applicants must be board certified/board eligible in Pediatric Cardiology or Pediatric Critical Care Medicine, or Pediatric Anesthesia and have either completed additional subspecialty advanced training or have specific experience in pediatric cardiovascular intensive care. Must also be licensed to practice medicine in the State of Louisiana by start date.

### Interested candidates should submit

a cover letter, CV and list of references electronically to  
[SOM-jobs@lsuhsc.edu](mailto:SOM-jobs@lsuhsc.edu).

Reference: Pediatric Cardiology Intensivist.

The School of Medicine does not participate in sponsoring faculty candidates for the Department of Health and Hospitals' Conrad 30 Program.

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### Global Heart Network Foundation (GHN)

a global non-profit organization with a mission to connect people and organizations focused on the delivery of cardiovascular care across the Globe to increase access to care.

Contact: [annabel@globalheartnetwork.net](mailto:annabel@globalheartnetwork.net)

[www.globalheartnetwork.net](http://www.globalheartnetwork.net)



## Florida Hospital for Children

### Pediatric Inpatient Cardiologist Orlando, Florida

The Walt Disney Pavilion at Florida Hospital for Children is looking for a Pediatric Cardiologist with primary inpatient care focus to help further develop our open heart program in Orlando. This physician will lead the cardiology portion of the pediatric cardiac intensive care team at Florida Hospital in Orlando as part of the multi-site John's Hopkins/All Children's Heart Surgery program.

Responsibilities of the new physician will include: cardiology management of all patients in our 10 bed pediatric CICU, hospital inpatient and NICU consults, post-op and post procedure management, performing TEEs and reading echocardiograms. Some outpatient clinic time is also possible depending on physician's level of interest. We have an active Extracorporeal Membrane Oxygenation (ECMO) Program, a pediatric residency program, and clinical research opportunities.

Working with an expanding and committed children's hospital, a collaborative relationship with Johns Hopkins and All Children's, an established and successful pediatric cardiology group with a strong referral base, a dedicated inpatient unit, a family-centered practice atmosphere, the ability to build and shape a program, and a competitive benefits / compensation package.

#### Requirements of the position:

- Diplomate (or board eligible) of the American Board of Pediatric Cardiology, extra training in cardiac intensive care is preferred.
- Experience and proficiency with TEE's and ECHO's.
- Strong leadership and communication skills.
- "Team player" mentality.

#### Contact:

Jason Junker  
Director, Physician Recruitment  
Phone: (407) 200-2538

[Jason.Junker@FLHosp.org](mailto:Jason.Junker@FLHosp.org)

<https://www.floridahospital.com/children>

patients. "The impact and advisability of that particular approach, while intuitively resonant, is still an open question. The arterial shunt at Stage II is yet another twist in the evolving story of hybrid therapy for HLHS." Dr. Nassar and co-investigators found that age and weight at second stage were lower in the arterial group than the venous group, suggesting that patients who had undergone the arterial shunt included those at higher risk. What is clear is that surgery for HLHS is an ongoing story as surgeons seek better outcomes with fewer complications for babies born with this serious abnormality.

### CONGENITAL CARDIOLOGY TODAY

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### HOW WE OPERATE



THE CONGENITAL HEART INTERVENTION  
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**Volunteer / Get Involved**  
[www.chimsupport.com](http://www.chimsupport.com)

The team involved at C.H.I.M.S. is largely a volunteering group of physicians nurses and technicians who are involved in caring for children with congenital heart disease.

The concept is straightforward. We are asking all interested catheter laboratories to register and donate surplus inventory which we will ship to help support CHD mission trips to developing countries.

# Interventional Pediatric Cardiologist



Pediatrix Cardiology is seeking a highly experienced board certified interventional pediatric cardiologist to serve as the Medical Director for The Pediatric Heart Center in Long Beach, CA, part of the Memorial Healthcare System — Miller Children’s and Women’s Hospital.

This position will partner with an existing team of three board certified pediatric cardiologists who provide an array of services including interventional, fetal, adult congenital heart disease, non-invasive EP and non-invasive imaging in addition to general pediatric cardiology services. The candidate will work collaboratively with the hospital’s cardiac surgeon to develop and promote this premier program throughout Southern California.

## Requirements:

- Leadership experience in a nationally recognized congenital heart center and/or large tertiary children’s hospital
- Experience in a state-of-the-art congenital cardiac cath lab
- Ability to assist the PICU team in developing a new state-of-the-art CVICU
- Knowledge of program development and fundraising
- Strong interpersonal skills with key healthcare and practice leadership

## Benefits:

- Located in desirable Southern California
- 93-bed Level 3 NICU and 24 additional private NICU bays, the largest in LA and San Diego, staffed by Pediatrix Medical Group/Mednax affiliated physicians
- Working collaboratively with the hospital cardiac surgeon
- Competitive salary and excellent benefits including health/dental/vision, 401(k), annual CME allowance, employee stock purchase plan, professional liability insurance, and assistance with mandatory hospital credentialing and state licensing

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**To apply for this position or learn about opportunities in other locations, please visit [www.pediatrix.com/clinicalcareers](http://www.pediatrix.com/clinicalcareers) or contact Janet Friedman, Physician Relations Specialist, Pediatrix Cardiology, at 800.243.3839, ext. 5589.**

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