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Surgical-Relevant Coronary Artery Anomalies Diagnosed by Cardiac Computed Tomographic Angiography in Infants and Children with Right Ventricular Outflow Tract Obstruction

By Erin E. Birmingham, MD; Abhineet M. Sharma, MD; Salil Ginde, MD; and Randy R. Richardson, MD

Key Words

Right Ventricular Outflow Tract Obstruction, Cardiac Computed Tomography Angiography, Coronary Artery Anomalies.

Introduction

Approximately 19% of congenital heart defects involve some degree of Right Ventricular Outflow Tract (RVOT) obstruction.¹ These obstructions can be at the level of the pulmonary valve, infundibulum, proximal branches, or distal branches of the pulmonary artery.² The severity of the clinical presentation is usually dependent on the severity of RVOT obstruction and can range from an asymptomatic infant to one that is severely cyanotic.

Many of these cases of RVOT obstruction mentioned above have associated Coronary Artery (CA) anomalies, known as prepulmonary coronary arteries. In this anomaly the coronary courses anterior to the pulmonary artery or RVOT.³ These prepulmonary coronary artery anomalies are associated with one or more cardiac malformations and alter surgical management if present.^{5,6} The prepulmonary

“Many of these cases of RVOT obstruction mentioned above have associated Coronary Artery (CA) anomalies, known as prepulmonary coronary arteries. In this anomaly the coronary courses anterior to the pulmonary artery or RVOT. These prepulmonary coronary artery anomalies are associated with one or more cardiac malformations and alter surgical management if present.^{5,6}”

coronary artery anomalies reviewed in this study include a prominent conal branch (Figure 1), a Left Coronary Artery (LCA) crossing in front of the RVOT (Figures 2, 3, 4), and a duplicated Left Anterior Descending Artery (LAD) (Figure 5). All of these anomalies can be inadvertently

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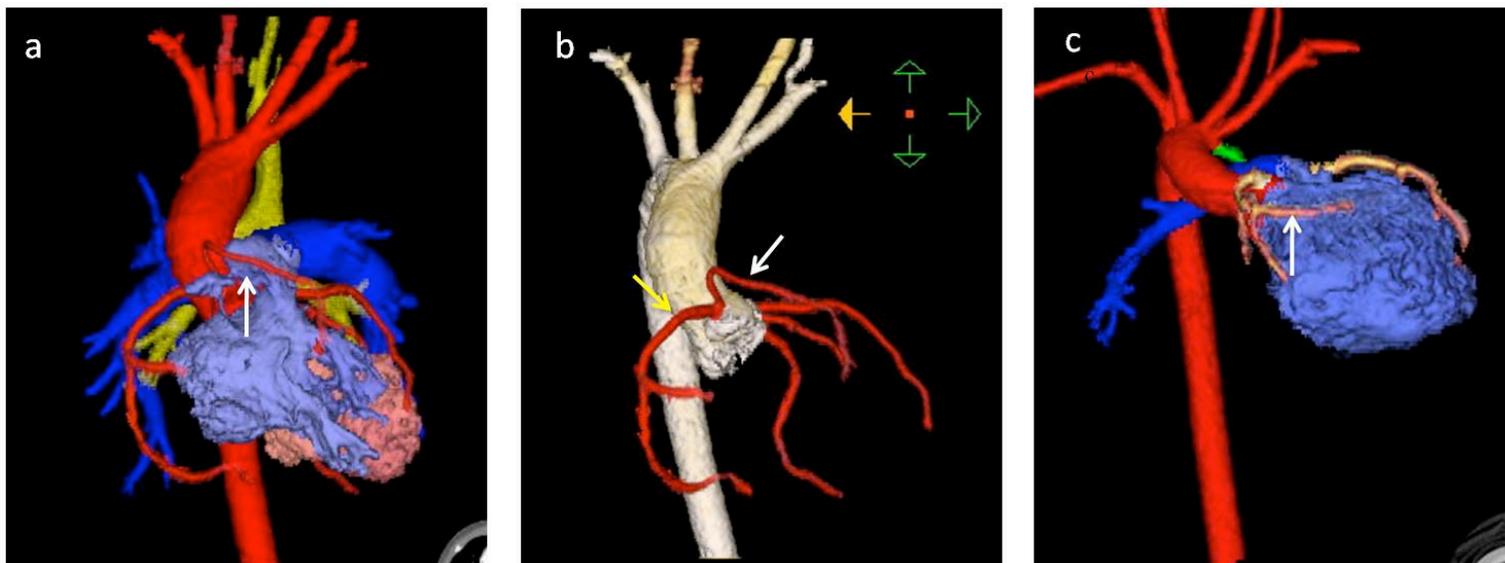


Figure 1. Prominent Conal Branch in a case of Tetralogy of Fallot (a, b) and Double-Outlet Right Ventricle (c). Color-coded 3D reconstructions. Anterior views of the heart showing the morphologic left ventricle (salmon), right ventricle (violet), aorta (a: red and b: neutral), and coronary arteries (red). In the case of Tetralogy of Fallot (a, b) The right coronary artery (yellow arrow) arises correctly from the right coronary sinus, however there is a prominent conal branch (white arrow) that arises off the RCA and courses anteriorly and slightly to the left. The left coronary artery has normal anatomy. This patient was found to have a constellation of findings compatible with TOF with subpulmonic pulmonary stenosis with post stenotic dilatation. In the case of DORV (c), the right coronary comes off the right coronary sinus as expected, however there is a prominent conal branch (white arrow) crossing over both the left and right outflow tracts.

damaged during surgical repair. In most of the cases of RVOT obstruction, the mainstay of treatment is interventional or intracardiac surgical management; however, the location of the CA anomalies can greatly affect what would otherwise be a routine surgical approach for a particular type of RVOT obstruction. Identifying these anomalies with the proper imaging modality prior to surgery can aid in the appropriate preoperative planning and avoid potentially life-threatening complications. The detection of coronary anomalies in children is challenging because they have smaller vessels, in addition to greater cardiac and respiratory motion compared with adult patients.⁷ When evaluating the coronary arteries in Congenital Heart Disease (CHD), it is most important to determine the exact origin, number, course, and termination of the arteries.³

Cardiac computed tomography with multidetector-row computed technology is a novel modality that can be used as a “one-stop shop” to non-invasively evaluate the coronary arteries, cardiac morphology, and cardiac function. It is also useful to assess coronary morphology and patency prior to and following surgical intervention for Congenital Heart Disease.⁸ Here, we compiled the largest number of coronary artery anomalies in RVOT obstruction evaluated by Coronary Computed Tomography Angiography (CCTA) in one study. Our study aimed to demonstrate the utility of CCTA as an imaging modality for the diagnosis of CA anomalies in cases of RVOT obstruction and CHD.

Materials and Methods

We retrospectively reviewed 220 standard preoperative CCTA exams in infants and

children performed over the period between 03/28/2005 and 09/18/2010 at St. Joseph’s Hospital and Medical Center in Phoenix, Arizona. All images were obtained as a part of preoperative protocol. Of the 220 exams, we found 67 patients with RVOT obstruction. Of the 67 patients, there were a total of 37 males and 30 females, ranging in age from 1 day to 15 years (Mean: 312 days; Median: 7 days). CCTA was performed with a 64-slice multi-detector CT, with prospective EKG gating, followed by three-dimensional reformations. Our institution’s current technique for CTA with 3D reconstruction uses prospective gating with padding to capture functional source data with less radiation. Images were captured from the thoracic inlet to the upper abdominal cavity. The clinically indicated 3D data set was acquired only once and was also used to assess the quality of the coronary images.

A Kruskal-Wallis analysis was used to see if there was a significant difference in the frequency of CA in the specific subtypes of RVOT. A Chi-squared test was utilized to see if there was a statistically significant difference in the frequency of CA anomalies in our RVOT population when compared to the overall population based on two studies.^{9, 10, 11}

The data analysis was approved by the institutional review board at St. Joseph’s Hospital and Medical Center. A waiver for informed consent was obtained. Literature was accessed through the PubMed search database using the phrases right ventricular outflow tract obstruction, coronary artery anomalies, cardiac computed tomographic angiography, and post-operative complications of Congenital Heart Disease.

No financial support was provided for data collection and analysis of manuscript preparation.

Results

Of the 220 CCTA exams performed, there were 67 patients with RVOT obstruction. These patients were grouped into 6 categories based on congenital heart anomaly (Table 1). Twenty-six of 67 patients were found to have Tetralogy of Fallot (TOF) and pulmonic stenosis (Group A). Fifteen patients had TOF and pulmonic atresia (Group B). Five patients had TOF with absent pulmonary valve (Group C). Four patients had isolated pulmonic stenosis (Group D). Seven patients had isolated pulmonic atresia (Group

Type of RVOT Obstruction	Percentage of Patients in Study Cohort (n)
Group A: Tetralogy of Fallot and Pulmonic Stenosis	38.8% (26)
Group B: TOF and Pulmonary Atresia	22.4% (15)
Group C: TOF with Absent Pulmonary Valve	7.5% (5)
Group D: Isolated Pulmonic Stenosis	6.0% (4)
Group E: Isolated Pulmonary Atresia	10.4% (7)
Group F: Double-Outlet Right Ventricle (DORV)	15.0% (10)
Total number of patients	67

Table 2. CA Anomalies Found in Cases of RVOT Obstruction

Group (n)	Prominent conal branch (n)	LCA in front of the RVOT (n)	Duplicated LAD (n)
A (7)	42.8% (6)	7% (1)	
B (2)	7% (1)		7% (1)
C (1)		7% (1)	
D (1)		7% (1)	
E (0)			
F (3)	7% (1)	14% (2)	
Total (14)	57% (8)	35.7% (5)	7% (1)

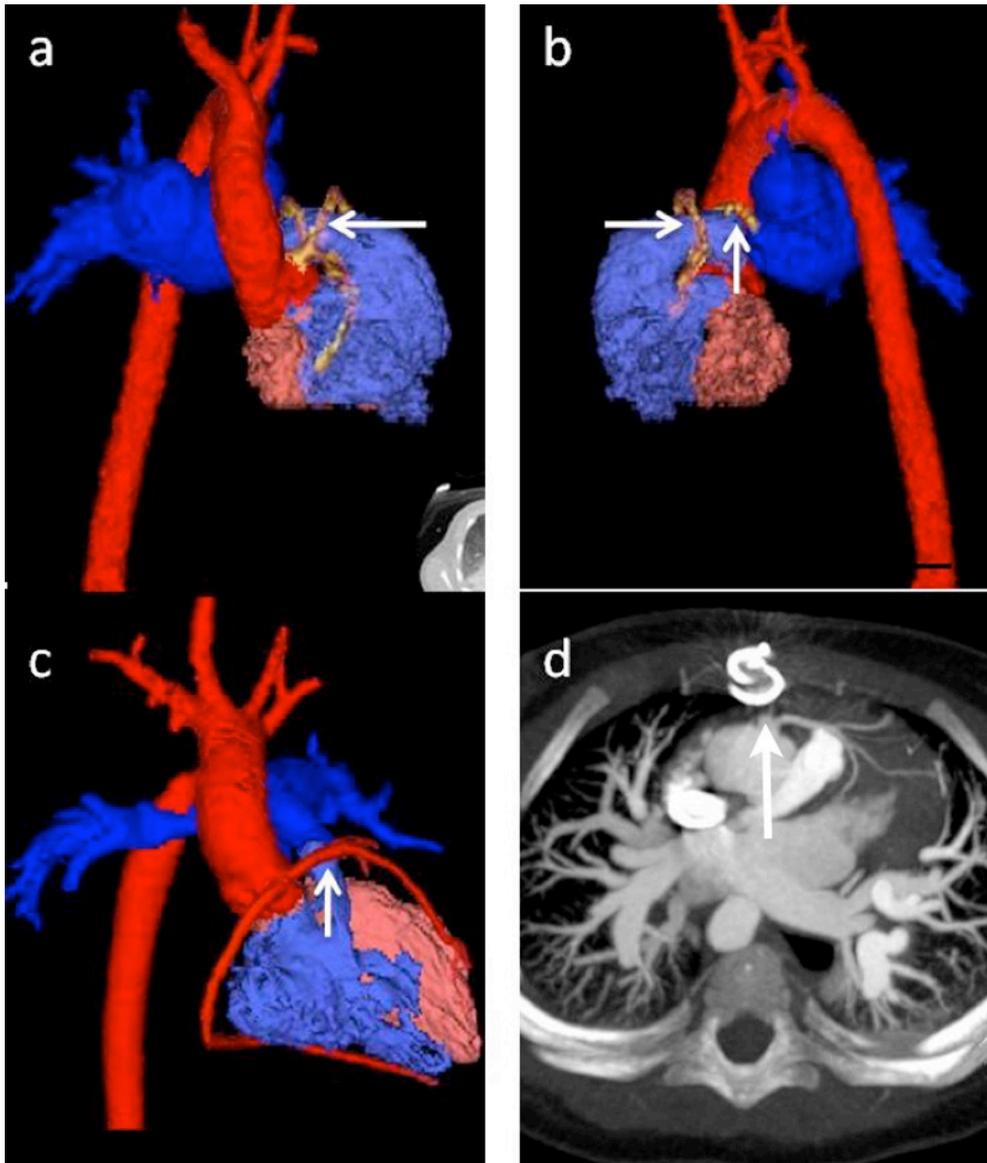


Figure 2. LCA in front of the RVOT in two cases of Tetralogy of Fallot. Color-coded 3D images showing the aorta (red), pulmonary arteries (dark blue), right ventricle (violet), left ventricle (salmon), coronary arteries (red and neutral).

Case 1: Anterior (a) and posterior (b) view of the entire heart. There is a overriding three-vessel left-sided aortic arch. The pulmonary valve is atretic with focal stenosis of the main pulmonary artery and massive post-stenotic dilatation of the right and left pulmonary arteries. A single coronary artery originates from the left sinus and trifurcates into the right, LAD, and circumflex arteries. The LAD and circumflex arteries (white arrows) have a malignant course anterior and superior to the right ventricular outflow tract. A subpulmonic VSD with right ventricular hypertrophy was also noted, consistent with findings of Tetralogy of Fallot.

Case 2: Anterior (c) and MIP images (d). There is a left coronary artery (white arrow) arising from the right coronary sinus which passes in front of the right ventricular outflow tract.

E). Ten patients had different forms of Double Outlet Right Ventricle (DORV) (Group F).

Fourteen of these 67 patients (20.9%) were found to have surgically relevant CA anomalies (Table 2). The most common CA anomaly seen with RVOT obstruction was the prominent conal branch, which made up 57% of cases. The prominent conal branch was seen most commonly in Tetralogy of Fallot with pulmonic stenosis. A Kruskal-Wallis analysis showed that there was no statistically significant association between RVOT subtype and frequency of coronary artery lesion ($H=1.86$, $df = 4$, $p = 0.76$). When compared to the incidence in the general population, the incidence of CA anomaly was higher in the RVOT Obstruction group when compared to studies done in the general population ($X^2 26.12$, $p = 0.0001$).

Discussion

Our results confirm that prominent coronary artery anomalies occur at a higher frequency in cases of RVOT obstruction when compared to the general population, with up to 20.6% of patients with RVOT obstruction having a CA anomaly. Additionally, our study found that CA anomalies do not occur more frequently in a specific type of RVOT obstruction, making it difficult to predict if surgeons might encounter a CA anomaly during a procedure. Our study shows that CCTA can be used to effectively diagnose various CA anomalies preoperatively and aid in operative management. For example, Figure 6 shows a case of a pre-pulmonary LAD artery that was found prior to surgery and altered surgical repair. Post-operative pictures show that the anomalous coronary artery was preserved without any post-operative complications. Hence, CA anomalies that would complicate the initial routine repair may require a different surgical approach.

Routine bedside echocardiography remains the first-line imaging modality to visualize the infant heart and coronary arteries due to its low cost, widespread availability, portability, ease of use, and excellent temporal-spatial resolution. As a result echocardiography reduces the need for diagnostic cardiac catheterization and angiography in most patients.^{8, 12} However, the diagnostic utility of echocardiography markedly diminishes with the growth of patients and after surgical procedures through median sternotomy because acoustic windows become progressively more limited.¹²

Previous studies have also reported the use of magnetic resonance imaging to diagnose various coronary artery anomalies in infants and children. Tangcharoen et al evaluated the feasibility and accuracy of magnetic resonance (MR) coronary angiography for the detection of CA anomalies in infants and children by using surgical findings as a

reference.⁷ Cardiac MRI offers several favorable characteristics in comparison with the conventional imaging modalities, including: good tissue characterization, being

operator independent, capacity for three-dimensional imaging, accurate flow measurements, ventricular function analysis, freely-selectable imaging planes, and lack of

ionizing radiation.¹² However, coronary MR angiography may be limited by irregular respiratory and cardiac rhythm, difficulty in delineating small CA's, and relatively long examination time.¹³ A comprehensive cardiac MRI investigation requires approximately 30 to 60 minutes with the patient in a still position in an uncomfortable environment; therefore, children under the age of 12 will commonly require anesthesia or conscious sedation.¹² As a result, it is often difficult to identify the complete coronary artery anatomy in most infants on MR, while CCTA can be a more reliable imaging modality in this age group.

CCTA is currently not the major modality for evaluating CA anomalies in Congenital Heart Disease, in particular RVOT obstruction. We found only one other study looking at coronary artery imaging with multislice CCTA. Hyun et al describes current multislice techniques for coronary artery imaging. The review was based on approximately 1,200 cardiac CT examinations over a period of 7 years. Hyun recommended that high temporal resolution, high z-axis spatial resolution, and effective suppression of cardiac motion are key CT parameters for visibility of CA's.¹³

Exposure to radiation remains a compelling concern when using CCTA; however, employing certain factors can minimize this risk. The imaging in our study was obtained using multi-detector row technology (64 multi-slice CT), to increase resolution and shorten the time spent imaging – allowing for

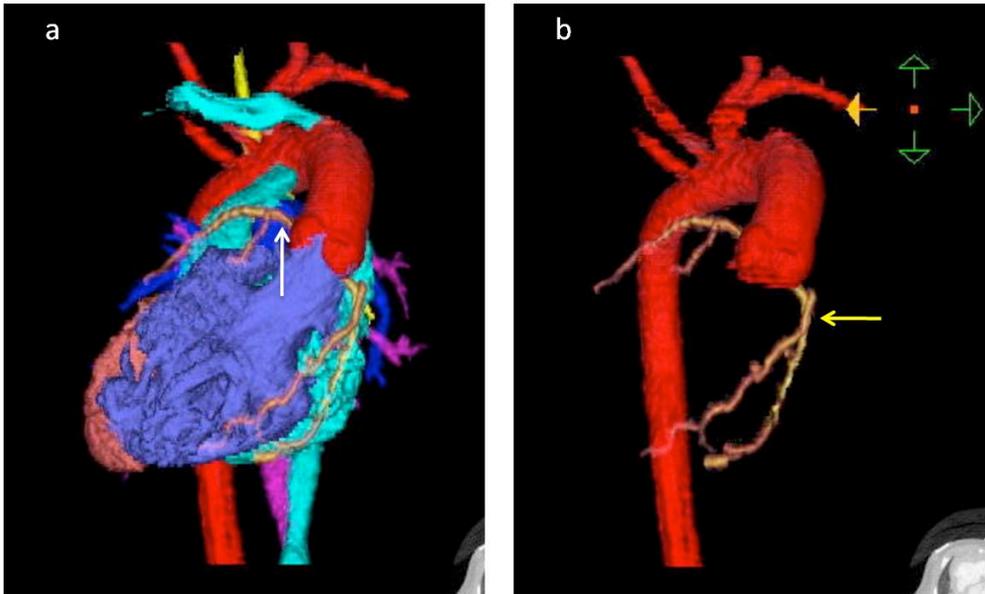


Figure 3. LCA in front of the RVOT in a case of Double-Outlet Right Ventricle. Color-coded 3D images. a. Anterior view of the entire heart. b. Anterior view of aorta (red) with coronary arteries (neutral). Both outflow tracts come off the morphologic right ventricle which is on the left and is enlarged (violet). There is a single coronary coming off the left coronary sinus. The morphologic left coronary artery which goes to the right goes between the right and left ventricular outflow tracts and is consistent with a malignant type of coronary artery course. There is an accessory left anterior descending artery coming off the morphologic right coronary artery, which is on the left and courses anterior to the right ventricle. This patient also has evidence of total anomalous pulmonary venous return, L-type Transverse Position of the Great Arteries, and heterotaxy with bilateral right-sidedness with thoracic situs inversus.

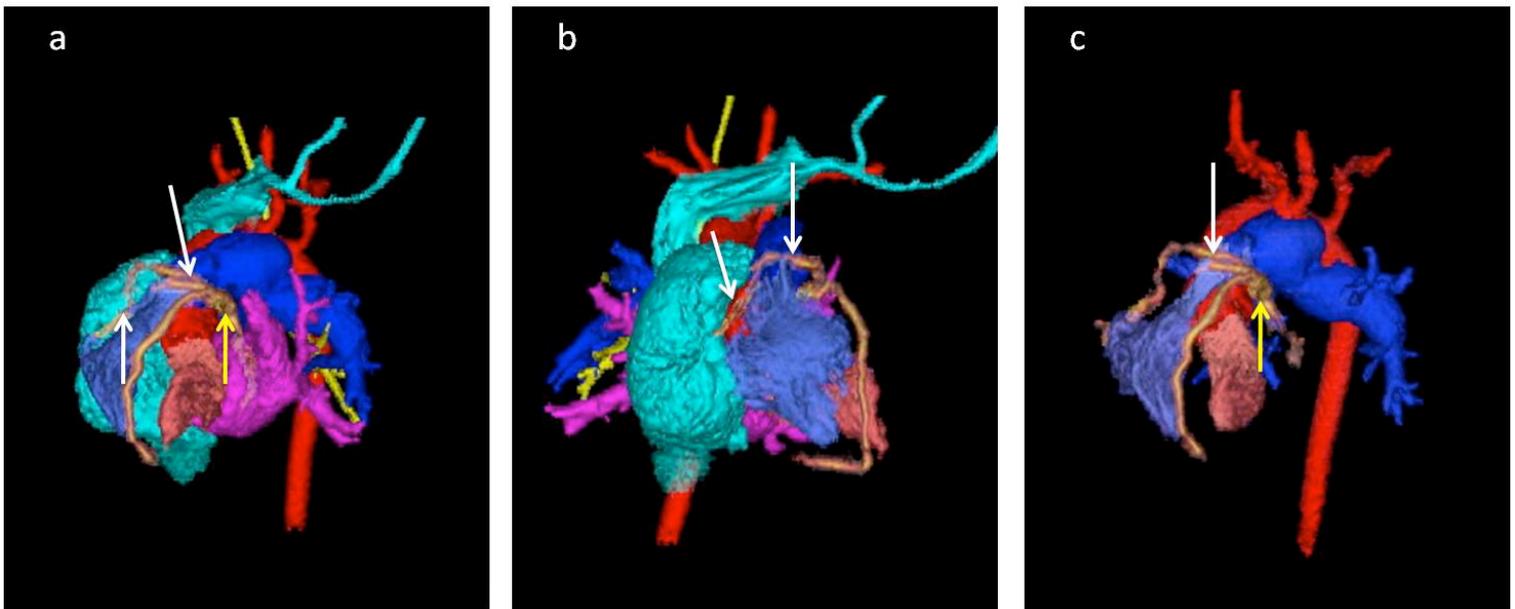


Figure 4. LCA in front of the RVOT in a case of dysplastic pulmonary valve and pulmonic stenosis: Color-coded 3D images. Anterior views (a and c) and posterior views (b) showing the aorta (red), pulmonary arteries (blue), left ventricle (salmon), right ventricle (violet), right atrium and SVC (light blue) and coronary arteries (neutral). The right coronary artery arises from the LAD (arrow) and courses anterior to the right ventricular outflow tract to the right atrioventricular groove. The left coronary artery originates from the left coronary sinus and is dilated (yellow arrow). Dysplastic pulmonary valves were found with stenosis of the pulmonary artery and right ventricular hypertrophy.

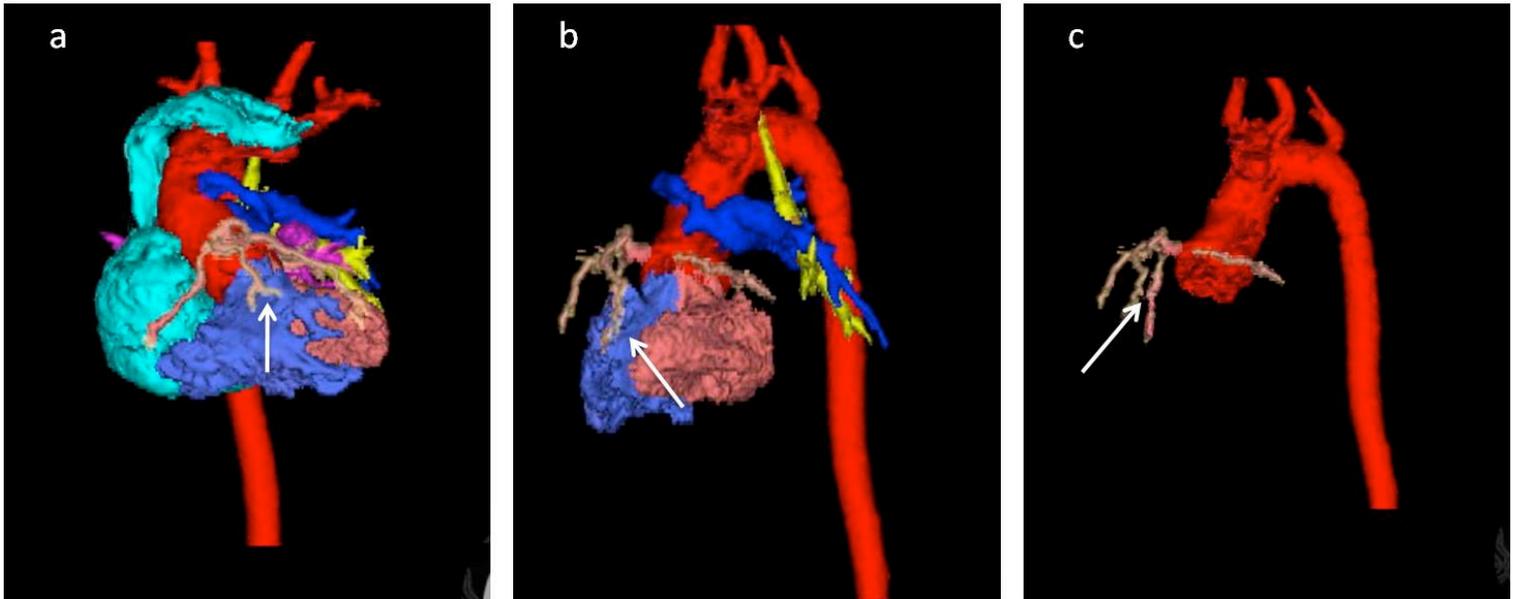


Figure 5. Duplicated LAD in Tetralogy of Fallot.

Color-coded 3D images. Anterior views of the heart showing the superior vena cava and right atrium (light blue), morphologic left ventricle (salmon), right ventricle (violet), aorta and coronary arteries (red). There is a three-vessel right-sided aortic arch with mirror imaging branching, an overriding aortic outlet, and rotated aortic root, which is common in Tetralogy of Fallot. The right coronary artery gives rise to an accessory left anterior descending artery (arrow) which courses anteriorly over the region of the expected pulmonary outflow tract. The left coronary artery arises normally and gives rise to an LAD as well (not shown). There is an atretic pulmonary outflow tract and a VSD consistent with findings of Tetralogy of Fallot.

less exposure to radiation. With multi-detector row CT, volumes of ionic IV contrast as small as 2 mL/kg allow successful contrast enhancement for CT angiography. Multislice CT has led to the ability to perform high-resolution scans gated to the cardiac cycle,¹² known as ECG-synchronized CT. These scans can be retrospectively gated or prospectively triggered. Strict breath-holding ability is necessary for retrospective ECG-gated CT in young children. Thus, prospective ECG-triggered CT is a viable option for free-breathing young children because the scan technique is less affected by respiratory motion.¹³ The use of ECG prospective gating in multislice CT scan allows a further reduction of the radiation exposure, thanks to the acquisition of images limited to 70-80% of the R-R interval.²⁰ In addition to reducing exposure to radiation, Hyun et al found that visibility of the coronary arteries by CT has markedly improved with prospectively ECG-synchronized CT.¹³

Due to the much shorter imaging time when compared to MR, the sedation rate for young children undergoing multidetector row CT is less than 5% [20]. Many of the studies in the literature support the idea that MR image quality and sharpness increased significantly with age and slower heart rates but did not perform as well in patients younger than four.⁷ However, CCTA image quality is independent of heart rates, which is very important in neonates with heart rates up to 160-180 beats per minute. Both Hyun et al and our study support the fact that high temporal resolution and ECG-synchronized data with CCTA improves visibility of the coronary

arteries and congenital heart defects beyond most other imaging studies.

This is the first study of its kind, investigating the use of CCTA to diagnose coronary artery anomalies in RVOT obstruction. Our intent was to establish a consistent imaging modality for the diagnosis of these surgically-relevant CA anomalies when echocardiography is equivocal and MRI is not feasible. This would allow surgeons to establish early and primary reparative surgery while maintaining the inherent CA anatomy. We have shown that CCTA with 3D reconstruction is an advanced, safe, and reliable imaging modality that provides accurate diagnosis even when echocardiographic data is unequivocal. The high-resolution 3D reconstructions are essential in the identification of the complex cardiac anatomy seen in these patients, and is necessary for preoperative planning, modified surgeries, and overall management of these patients.

Conclusion

Many cases of RVOT obstruction have associated coronary artery (CA) anomalies that can go undiagnosed prior to surgery. Multidetector cardiac CTA with EKG gating is a safe and feasible tool for the detection of surgically-relevant CA anomalies in patients with RVOT stenosis or obstruction. Abnormal CA anatomy must be considered prior to surgery in any infant with CHD and intracardiac defects. CCTA is very useful adjunct imaging modality for preoperative planning and overall management, when Echocardiographic imaging is unclear.

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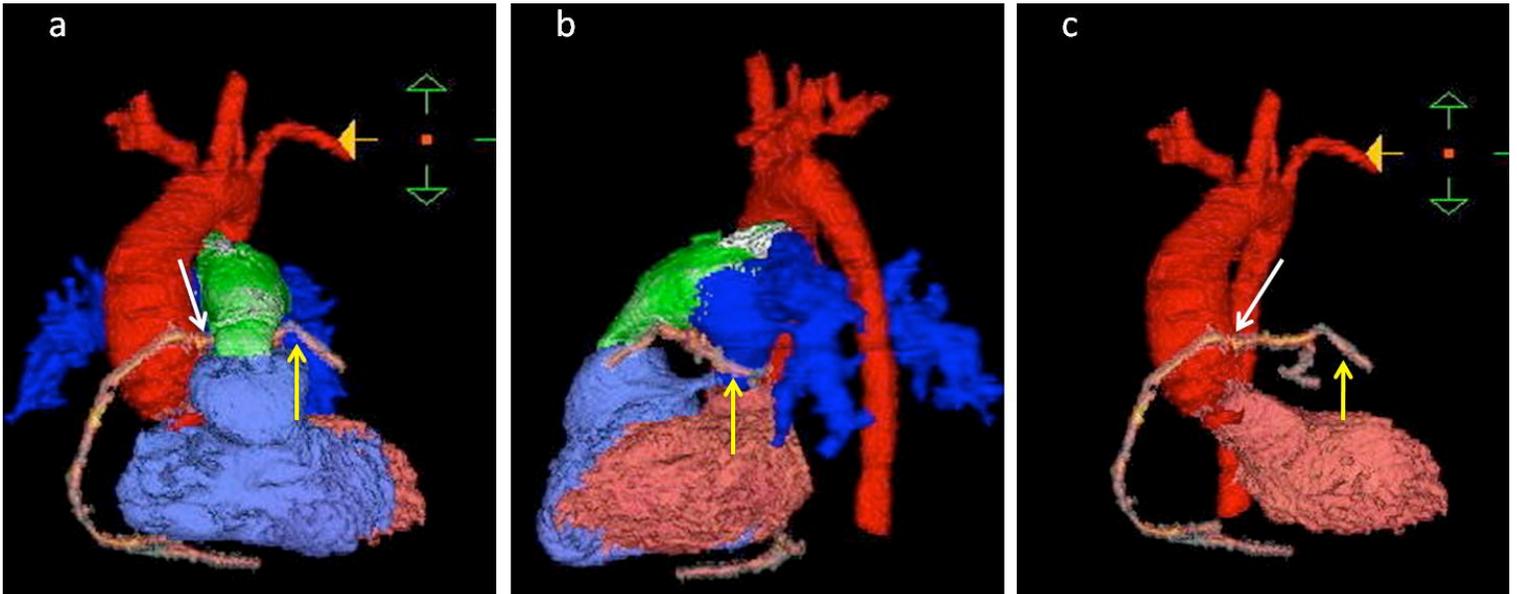


Figure 6. Post-operative images showing preservation of the anomalous CA. Color-coded 3D images. Anterior views (a and c) and posterior view (b) of the entire heart. Showing the left ventricle (salmon), right ventricle (violet), aorta (red), pulmonary arteries (blue), surgically repaired pulmonary outflow tract (green), and coronary arteries (neutral). This patient had a single LCA (white arrow) with a prepulmonary LAD artery (yellow arrow) that was found prior to surgery and altered surgical repair. These post-operative pictures show that the anomalous coronary artery was able to be preserved without any post-operative complications.

Limitations

Limitations to this study include the retrospective study design and that there was no direct comparison between CCTA and other imaging modalities. In addition, it is unclear if the coronary anomalies seen on CT imaging were confirmed at the time of surgery.

Compliance with Ethical Standards

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research board and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. For this type of study formal consent was not required.

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“Many cases of RVOT obstruction have associated coronary artery (CA) anomalies that can go undiagnosed prior to surgery. Multidetector cardiac CTA with EKG gating is a safe and feasible tool for the detection of surgically-relevant CA anomalies in patients with RVOT stenosis or obstruction. Abnormal CA anatomy must be considered prior to surgery in any infant with CHD and intracardiac defects. CCTA is very useful adjunct imaging modality for preoperative planning and overall management, when Echocardiographic imaging is unclear.”

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Biographical Sketch

Erin Birmingham, MD is currently in her second year of pediatrics residency at Children's Hospital of Wisconsin/Medical College of Wisconsin. She obtained her medical degree from Creighton University School of Medicine.

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Compiled and Reviewed by Tony Carlson, Senior Editor

The 7th World Congress of Pediatric Cardiology & Cardiac Surgery Has Changed Date and Venue: July 16th-21st, 2017, Barcelona, Spain

The Organizing Committee has announced the 7th World Congress of Pediatric Cardiology and Cardiac Surgery (WCPCCS), will take place on July 16th - 21st, 2017, in the Centre Convencions Internacional de Barcelona (CCIB), Barcelona, Spain.

“The 7th World Congress of Pediatric Cardiology & Cardiac Surgery has changed date and venue: July 16th-21st, 2017, Barcelona, Spain”

The aim of WCPCCS is to bring together all professionals involved in the care of Pediatric Heart Disease and Congenital Heart Disease of all ages, from the fetus to the aged. The Congress will provide a unique opportunity to meet the leaders of specialties worldwide; to learn about the latest innovations and the results of procedures; and to contribute to the discussions, debates and plenary sessions with renowned speakers.

The central philosophy of the Congress is “bridging together” all major specialties in the field. Accordingly, the scientific program is carefully planned to address all interests and expertise with concentration streams on pediatric cardiology, pediatric cardiac surgery, adult congenital heart diseases, anesthesia, intensive care and nursing.

We are excited to offer the scientific and cultural feast of a lifetime to one of the most refined crowds in the profession, in one the most welcoming, inimitably exciting venues of the world. For more information visit: www.wcpccs2017.org.

Study Identifies a Developmental Cause of Cardiac Hypertrophy - New Findings Provide Important Insights into a Leading Cause of Heart Failure

Newswise – Investigators at Beth Israel Deaconess Medical Center (BIDMC) have identified a developmental cause of adult-onset cardiac hypertrophy, a dangerous thickening of the heart muscle that can lead to heart failure and death. Reported July 7th online in *The Journal of Clinical Investigation*, the new findings could lead to targeted therapies for this condition.

“Genetic mutations that alter signaling pathways involved in cardiac development have been implicated in approximately 30% of birth defects associated with Congenital Heart Disease,” explained Maria Kontaridis, PhD, Interim Director of Basic Cardiovascular Research in the CardioVascular Institute at BIDMC and Assistant Professor of Medicine at Harvard Medical School. “In this study, we wanted to determine whether these same genetic mutations can also lead to the development of cardiac hypertrophy in adults.”

Work in the Kontaridis lab focuses on a cluster of genetic diseases known as RASopathies, which impact the Ras/MAPK cell signaling pathway and affect the growth and differentiation of cells. One of these diseases, Noonan Syndrome with Multiple Lentigines (NSML), is a rare condition caused primarily by mutations in the gene PTPN11. NSML has multiple phenotypic characteristics, including short stature, growth retardation, and craniofacial abnormalities, but the most prevalent defects affect the heart, the most common of which is cardiac hypertrophy.

By creating NSML mice that expressed the PTPN11 mutation, the scientists were able to gain important insights into what causes Congenital Heart Disease and how these defects lead to hypertrophy.

“In this study, we observed that there were significant developmental abnormalities in hearts affected by NSML,” explained Kontaridis. “We wanted to figure out if the adult-onset cardiac hypertrophy we observed might be caused by abnormal regulation of one or more of the critical cell populations necessary for growth and development of the heart.”

Cardiac development consists of a sequential order of events that are controlled by intercellular communication between three critical cell populations, which become the heart’s endocardium, myocardium and neural crest regions. Carefully orchestrated cross-talk between these cell populations culminates in a fully functional heart. Multiple cell signaling pathways regulate these complex developmental events, but when these pathways are disrupted, communication breaks down, leading to various cardiac defects.

“Our experiments unexpectedly revealed that hypertrophy was caused by abnormal cell signaling mechanisms originating from developing endocardium—not the myocardium, as had been long assumed,” said Kontaridis.

Experiments with mice expressing the genetic mutation only in the cells destined to give rise to the endocardium showed that as these mice aged, they developed hypertrophic cardiomyopathy. Mice with mutations in the myocardial- or neural crest- regions did not show signs of associated hypertrophy, definitively identifying the endocardium as the source of hypertrophy in NSML.

“This was the first time we were able to trace the cause of an adult-onset hypertrophic disease to abnormal development, and



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specifically to a disruption in critical endocardial to myocardial cell 'crosstalk,'" explained Kontaridis.

"These findings not only tell us that there are developmental components to NSML that we can potentially target therapeutically, but also suggest that we can start to think about what the signaling differences may be between this RASopathy and other RASopathy disorders that cause similar, but different cardiac defects," said Kontaridis. "More important, we can consider these same approaches when thinking about treating other, more common, congenital hypertrophy disorders."

Study coauthors include: BIDMC investigators Jessica Lauriol (first author), Janel R. Cabrera, Ashbeel Roy, Kimberly Keith, Sara M. Hough, Federico Damilano, Bonnie Wang and Saumya Das; Roderick Bronson of Harvard Medical School; and Gabriel C. Segarra, Meaghan E. Flessa, Lauren E. Miller and Kyu-Ho Lee of the Medical University of South Carolina

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For more information, visit www.bidmc.org.

Correction to August 2016 Issue:

The August 2016 article, "Double-Orifice Tricuspid Valve: Case Report with a Review," by DR Hakim Irfan Showkat, MBCh, MD; Rekha Mishra, MD; Vinod Sharma, MD, DM; Lokesh Chandra Gupta, MD, DM; and Sadaf Anwar, PGDCC, had some missing author information. It is correct above, and has been corrected on the PDF versions, which can be found on page 7 for both the North American and International editions at:

North American Edition
www.congenitalcardiologytoday.com/index_files/CCT-AUG16-NA.pdf

International Edition
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10th Biennial 11q Research and Resource Conference - June 26th-30th, 2016, San Diego, California

By Paul Grossfeld, MD

In July, 1995 during my first month of Pediatric Cardiology fellowship at the University of California San Diego (UCSD), I took care of a patient with Jacobsen Syndrome (JS), a rare chromosomal disorder caused by deletions in the long arm of chromosome 11. My interest in this syndrome was heightened by the possibility of identifying a novel gene for left-sided obstructive lesions, including HLHS (Hypoplastic Left Heart Syndrome), which we subsequently discovered occurs at a higher frequency in JS than for any other known human genetic syndrome. Fast forward to June, 2016: serving as the Chief Medical Advisor for the past 15 years for the *11q Research and Resource Group* (www.11qusa.org), a support group for patients and their families with JS, I worked with the group's President, Linzee Carroll (mother of two children with JS), to organize the *10th Biennial International Conference* held in San Diego, June 26th-30th. Fifty-one families and 15 invited faculty from nine countries representing four continents, gathered for the four-day conference. Just over half of the families were attending for the first time.

While my initial interest was and continues to be in the genetic basis of HLHS and other congenital heart defects that occur in JS, through the encouragement of the families I have established collaborations that have focused on many of the other major causes of morbidity and mortality in JS including: a congenital platelet disorder, varying degrees of intellectual disability, behavioral problems including autism and ADHD (Attention Deficit Hyperactivity Disorder), and life-threatening immunodeficiencies.

Day One of the conference began with the keynote speaker, Ms. Peyton Goddard (www.peytongoddard.com), who depicted her struggles as a severely autistic and non-verbal child. She described how using facilitated communication allowed her to overcome her limitations, as well as the discrimination, abuses and biases of others to accomplish, among her many remarkable achievements, graduating valedictorian from her college. The remainder of the day included talks on several of the recently-identified major medical issues including immunodeficiency and brain hemorrhages. The afternoon featured a



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riveting talk by Reverend Ryan Sey, the head chaplain at Rady Children's Hospital of San Diego, on coping with having a child with a rare genetic disorder. The opening evening dinner featured every child receiving a "rare bear," graciously provided by Dr. Chris Waters from her non-profit organization, Rare Science (www.rarescience.org). That was followed by a talent show displaying the remarkable talents of several of the JS children and young adults.

Tuesday's sessions were highlighted by several talks on neurologic and behavioral problems, including a hands-on demonstration by chiropractor Dr. Tommy John III on how improving movement can help overcome some of the physical limitations imposed by the syndrome. The afternoon session included a lively panel discussion on potential gene-based therapies for intellectual disability and autism, with the hopes of launching at least one pilot clinical trial by the end of 2016. The day culminated by attending the San Diego Padres game that evening, with tickets graciously donated for all of the families by the Padres.

Wednesday began with a comprehensive overview of endocrine problems that can occur in JS, followed by a number of breakout sessions led mostly by family members of affected children. Perhaps the highlight and most moving session of the conference was the Wednesday afternoon panel discussion consisting of adults with JS, as they fielded questions about what it is like to live with JS. The remainder of the afternoon consisted of more breakout sessions, including another gripping discussion led by Reverend Sey entitled, "Shut Up About Your Perfect Child."

Thursday, the final day of the conference, was highlighted by an interactive music therapy session organized by Dr. Barbara Reuer and three of her music therapists (www.resoundingjoyinc.org). The emphasis

was on participation, and people of all ages with JS found themselves engaged and thoroughly enjoying the moment. The conference concluded with a picnic in which families had one final opportunity to say goodbye to each other until the next conference, tentatively scheduled for the summer of 2018.

Numerous on-site assessments took place during the conference, including: cognitive and behavioral, three-dimensional craniofacial imaging, physical exams, and one-on-one family meetings with me. These assessments have been instrumental in acquiring new knowledge, ultimately leading to improved therapies for people with JS.

Financial support was provided, in part, by the Jeffrey Modell Foundation (jmfworld.com), Vivint Gives Back (www.vivint.com/company/gives-back), and the UCSD Department of Pediatrics (www.pediatrics.ucsd.edu). The Sulpizio family kindly donated Magali wine for all of the speakers. In addition, many families did their own fundraising to afford the cost of attending the conference, and thanks to some generous anonymous donors, limited scholarships were made available to families in financial need. In one case the father of a child from England, who sold his guitar in order to afford the costs of bringing his family to the conference, was given a brand new handmade guitar graciously provided by Pepe Romero Jr., of the famous Romeros family of classical guitarists.

Ironically, it was my interest in the broken hearts of people with JS that initially got me involved with this remarkable group of families. To that end, we have made some exciting strides in identifying the gene that causes congenital heart defects in JS, a second genetic locus that can prevent heart defects in mice lacking the gene that causes heart defects, and most recently an animal model for HLHS using the commonly studied frog *Xenopus*. Despite these very gratifying successes, I never anticipated that 20 years after seeing my first JS patient, it would be the strength and beauty of their hearts that could teach me so much about the human condition, and why I remain involved with this unique and extraordinary group of families.

CCT

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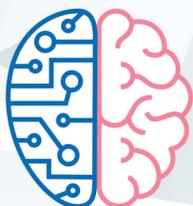
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