

# CONGENITAL CARDIOLOGY TODAY

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(See [www.cct.bz](http://www.cct.bz) for additional meetings)

#### 5<sup>th</sup> Phoenix Fetal Cardiology Symposium

Apr. 23-27, 2014; Phoenix, AZ USA  
[kpoole@phoenixchildrens.com](mailto:kpoole@phoenixchildrens.com)

#### The 61<sup>st</sup> Annual Conference of the Israel Heart Society in association with the Israel Society of Cardiothoracic Surgery

Apr. 30-May1, 2014; Tel-Aviv, Israel  
[israelheart.com](http://israelheart.com)

#### World Congress of Cardiology Scientific Sessions 2014

May 4-7, 2014; Melbourne, Australia  
[www.worldcardiocongress.org](http://www.worldcardiocongress.org)

#### 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/>

#### Cardiac Imaging for Structural Heart Disease Special One Day Symposium at PICS-AICS

Jun. 7, 2014; Chicago, IL USA  
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## Real-Time-Three-Dimensional Echocardiographic Assessment of Atrial Septal Defects

By Donald J. Hagler, MD

Previous reports have demonstrated the utility of real-time three-dimensional echocardiographic (RT3DE) imaging in the assessment of atrial septal defects (ASDs) prior to both surgical and device closure interventions.<sup>1-4</sup> To date, the reports have illustrated that secundum ASDs may have normal shape and location (Figure 1), or unusual oblong or irregular shapes, and may be multiple (Figures 2 and 3). RT3DE also has been particularly useful for illustrating the surrounding rims of the secundum ASD, and the abnormal location and absent rims of sinus venosus type defects. The purpose of this report is to extend these initial observations to include RT3DE Transesophageal (TEE) imaging of atrio-ventricular (AV) septal defects, and to review some preliminary experiences with RT3DE intracardiac (ICE) imaging of ASD prior to device closure.

Recent reports of RT3DE ICE imaging discussed the use of Acu Nav V 3-dimensional ultrasound catheter and SC2000 imaging platform (Siemens Healthcare, Mountain View, CA) during interventional electrophysiologic procedures.<sup>5</sup> The AcuNav V catheter is a 90 cm 10-French phase array probe similar to the 2D AcuNav catheter capable of articulating 160 degrees in four directions. It provides real-time (4D) imaging with a volume size of 22 degrees by 90 degrees with a variable, multi-MHz frequency capability. The report illustrates the advantage of volume imaging in providing visualization of the entire pulmonary vein ostium.

Intraoperative TEE has been the standard of practice for assessment of patients with more complex forms of congenital heart disease. The addition of RT3DE in the TEE assessment provides views which allow the surgeon to view the cardiac structures in a format similar to the anatomic views obtained during cardiopulmonary bypass. Figures 4A and B illustrate diastolic and systolic 3D images of an adult patient with a partial form of AV septal defect and a common atrium. The diastolic view focuses on the crest of the ventricular septum (VS) with no evidence of intervening atrial septum. The systolic frame

***“Intraoperative TEE has been the standard of practice for assessment of patients with more complex forms of congenital heart disease. The addition of RT3DE in the TEE assessment provides views which allow the surgeon to view the cardiac structures in a format similar to the anatomic views obtained during cardiopulmonary bypass.”***

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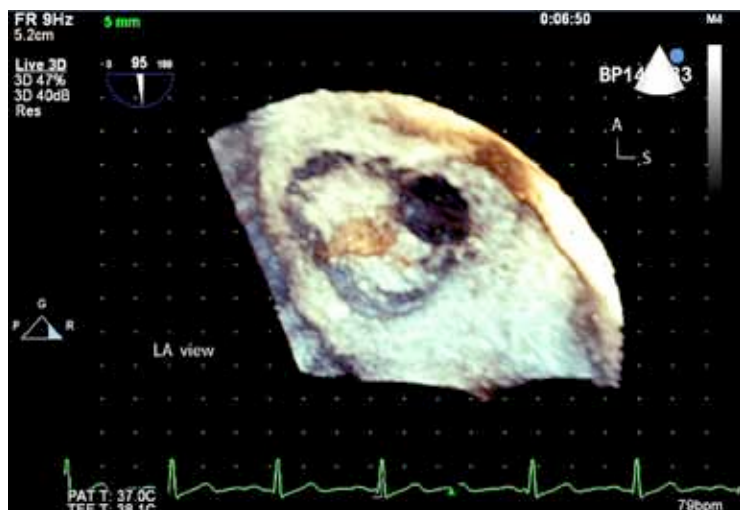


Figure 1. 3D Transesophageal (TEE) still frame volume image of a moderate sized ostium secundum atrial septal defect view from the left atrial aspect. The defect is round, symmetrical and located just below the limbus of the fossa ovalis. The remaining portion of the septum primum is intact.

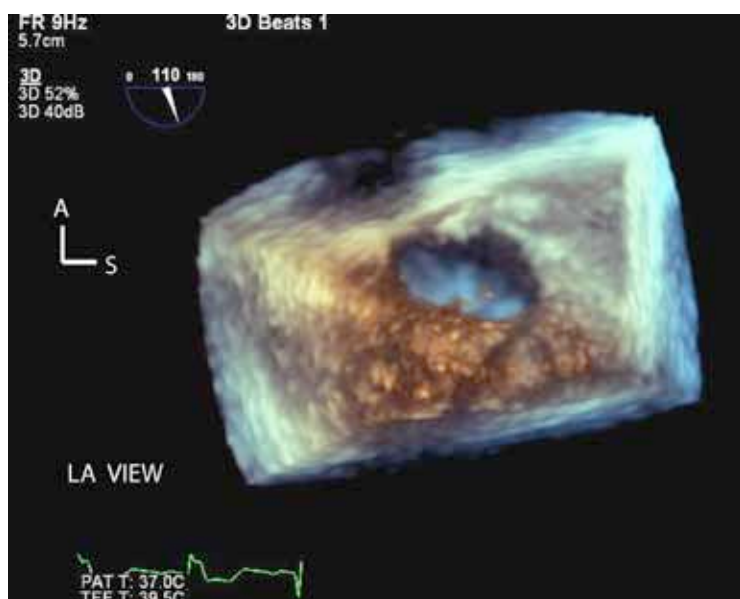


Figure 2. 3D TEE still frame volume image of a large, fairly oblong and irregularly shaped secundum ASD located more inferiorly in the atrial septum. No other defects are evident.

demonstrates effective closure of both AV valve with the line of closure of the cleft anterior mitral leaflet evident with real-time imaging. The post-operative image (Figure 4C) illustrates effective repair of the cleft in the anterior mitral leaflet observed in a short axis format. The valve was completely competent after surgical repair of the mitral valve cleft.



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The symposium will include sessions that focus on the latest updates in prenatal therapies and fetal cardiac and non-cardiac interventions, as well as new fetal imaging modalities. Attendees will have multiple opportunities to participate in hands-on workshops under the supervision of expert faculty in the field of fetal cardiology. There will also be opportunities to discuss interesting cases along with Q&A sessions.

### Course Directors:

Sawsan M. Awad, MD, MSC; Karim Diab, MD, FACC, FASE

### Course Co-Directors:

Emerio T. Alboliras, MD, FAAP, FACC, FASE; John Bokowski, PhD, RDCS, FA; Xavier Pombar, DO; Mark Sklansky, MD

### Invited National and International Faculty:

In addition to the above there will be: Lisa Hornberger, MD; Edgar Jaeggi, MD, FRCP(C); Anthony Johnson, DO; Carlos AC Pedra, MD, PhD; Simone Fontes Pedra, MD; Ra-id Abdulla, MD; Sawsan M. Awad, MD, MSC; John Bokowski, PhD, RDCS, FASE; Xavier Pombar, DO; Debra Selip, MD, FAAP; Michelle Rexilius, RDCS

### Overview of Some of the Selected Topics at the Meeting:

- Fetal Cardiac Screening: basic views and red flags
- Live Scanning: Step-by-Step Approach for Complete Fetal Echocardiography Study
- Practical Tips in Diagnosing Anomalies of the 4-Chamber View (TAPVR, AVC, Ebstein, ASD, VSD)
- Practical Tips in Diagnosing Anomalies of the 4-Chamber View (TA, MA/ HLHS, DILV)
- Practical Tips in Diagnosing Anomalies of the Outflow Tracts (Truncus arteriosus/DORV/TOF-PA/Absent Pulmonary Valve)
- Practical Tips in Diagnosing Anomalies of the Outflow Tracts (TGA, AS, PS)
- Coarctation of the Aorta/ Interrupted Aortic Arch/ Vascular Rings
- Heterotaxy Syndrome and VisceroAtrial Situs Abnormalities
- Abnormalities of the Fetal Patent Ductus Arteriosus
- Borderline Common Cardiac Findings in the Fetus
- Cardiovascular Physiology: Pathophysiological Changes of CHD
- Fetal Arrhythmias: Diagnosis and Management
- How to Start a Fetal Cardiac Intervention Program
- Fetal Cardiac Intervention: Current and Future Directions
- Twin Twin Transfusion Syndrome: Cardiac Compromise and In Utero Treatment
- Fetal Intervention Ethics
- Fetal Cardiac Cases
- How to Evaluate Fetal Cardiac Function: Standard & New Modalities
- Recognition of Atrial Septal Restriction in Complex Congenital Heart Disease

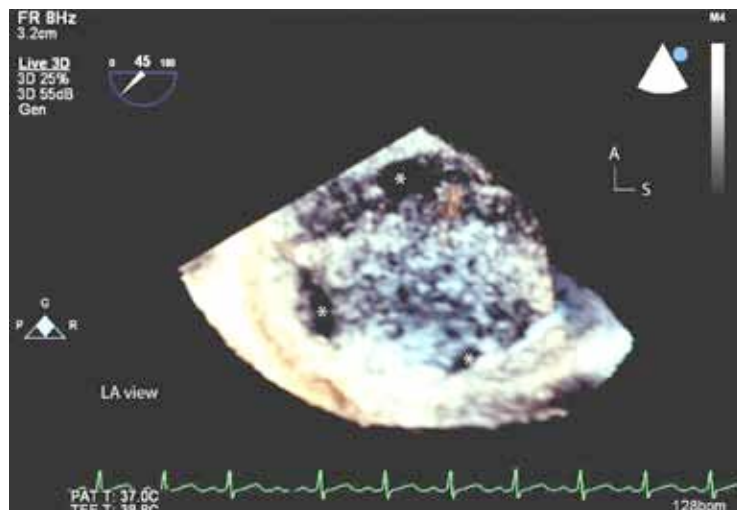


Figure 3. 3D TEE still frame volume image viewed from the left atrium demonstrating multiple (3) ostium secundum defects (asterisks) located in separate and isolated segments of the atrial septum. The largest defect is more anterior and below the limbus of the fossa ovalis.

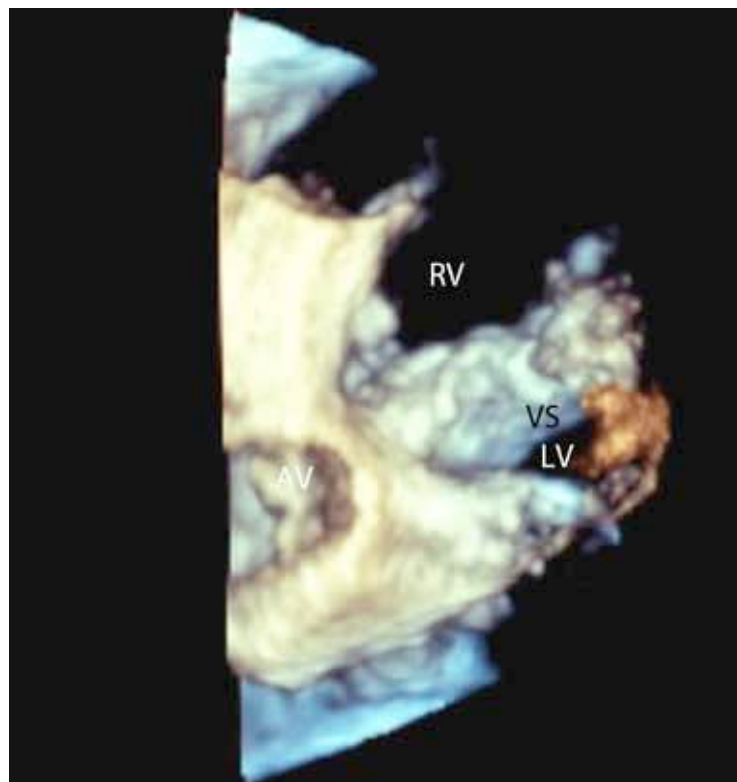


Figure 4A. 3D intraoperative TEE still frame volume image demonstrating a large ostium primum atrial septal defect (common atrium). A diastolic frame looking down on the crest of the ventricular septum (VS) through both atrioventricular valve (AV) orifices.

LIVE CASES

**JUNE 26 – 28, 2014**  
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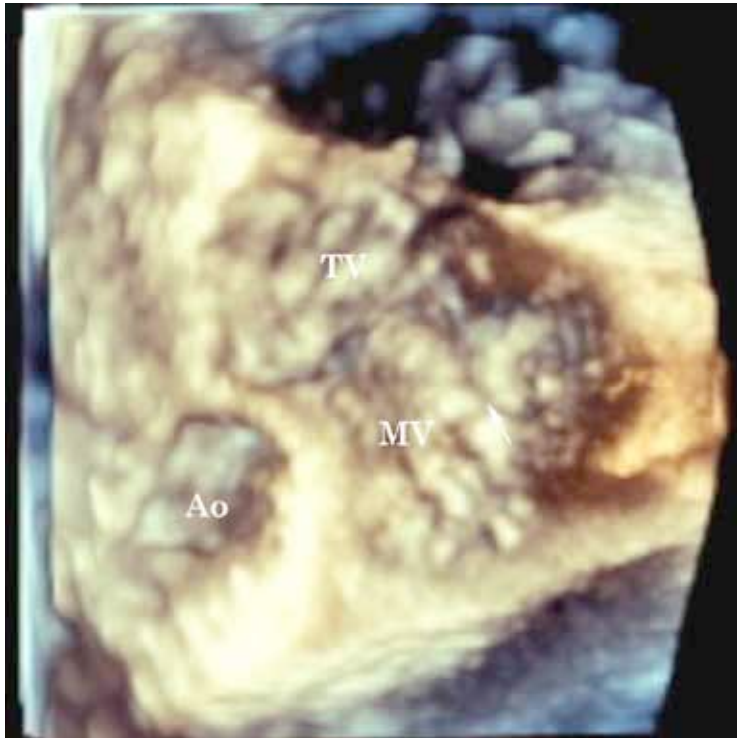


Figure 4B. A systolic frame from the same projection demonstrating closure of both AV valves. The arrow points to the coaptation point of the cleft in the anterior mitral leaflet.

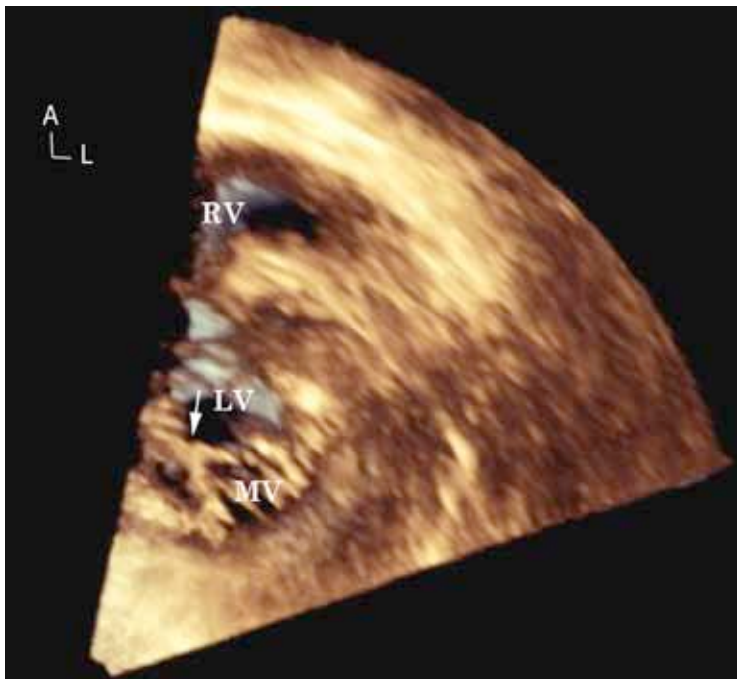


Figure 4C. A 3D still frame image illustrating a cross sectional view of the mitral valve orifice after surgical repair. The arrow points to the area of repair of the cleft in the anterior mitral leaflet.

Concordant with the observations of improved recognition of the structural relations of atrial septal defect observed with 3D TEE imaging of secundum defects during surgical or interventional device closure procedure, we have utilized RT3DE ICE imaging in our assessment of patients during procedures for interventional device closure. Figures 5A, B and C illustrate typical features of a recurrent secundum atrial septal defect after attempted surgical repair. Figure

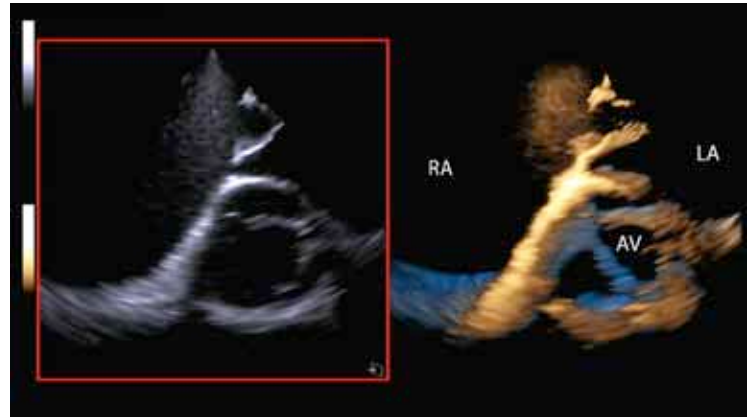


Figure 5A. 2D and 3D comparative images of a recurrent secundum atrial septal defect obtained with 3D ICE imaging.

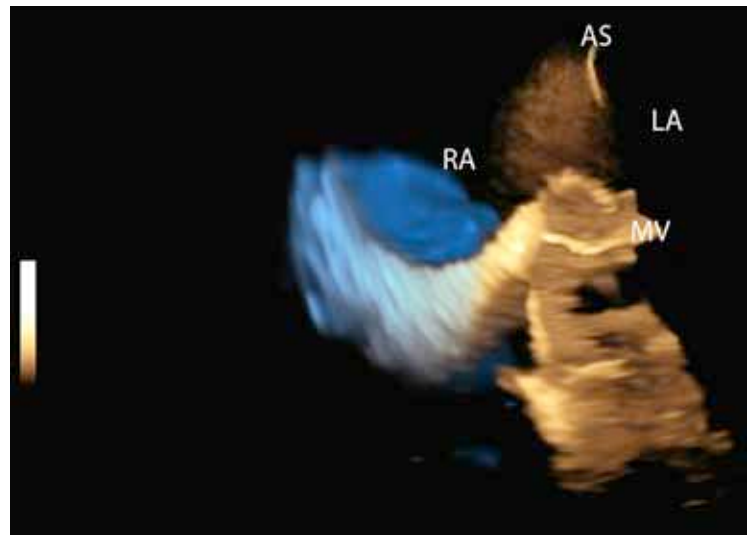


Figure 5B. Rotated 3D ICE image illustrating the left atrial aspect of the recurrent atrial septal defect and its relationship to the mitral valve just inferior to the defect.

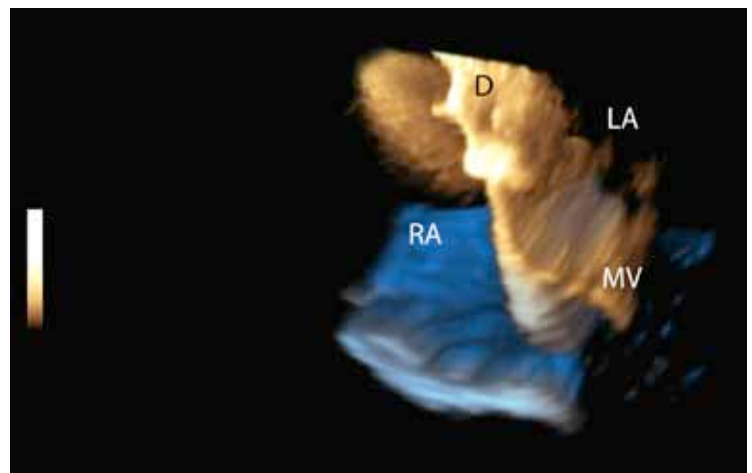


Figure 5C. Similarly rotated 3D ICE image demonstrating the left atrial aspect of an Amplatz atrial septal occluder closing the recurrent defect. The device does not interfere with the mitral valve function.

5A demonstrates a comparative image of 2D and 3D images obtained simultaneously with the AcuNav V catheter. Multiple defects were evident in close proximity with flail segments of disrupted tissue used in the surgical repair. Figure 5B illustrates the utility of RT3DE ICE



images to allow rotation of the 3D image to visualize the left atrial (LA) aspect of the recurrent defect and its relationship to the mitral valve (MV) observed inferiorly. Figure 5C illustrates the closed defect with an Amplatzer atrial septal occluder (St. Jude Medical, St. Paul, MN) with no evidence of residual defect and no interference with the mitral valve.

Although the RT3DE ICE images are limited with the 20 degree format currently available, they provide additional imaging capability and view of cardiac structures not possible with standard 2D ICE images. This topic will be discussed in more detail and with additional demonstration in video format at the 22<sup>nd</sup> Annual International Echo Symposium in Parma, Italy, June 16<sup>th</sup>-18<sup>th</sup>, 2014.

***“Although the RT3DE ICE images are limited with the 20 degree format currently available, they provide additional imaging capability and view of cardiac structures not possible with standard 2D ICE images.”***

## References

1. Marx GR, Fulton DR, Pandian NG, Vogel M, Cao QL, Ludomirsky A, et al. Delineation of site, relative size and dynamic geometry of atrial septal defect by real-time three-dimensional echocardiography. J Am Coll Cardiol 1995;25: 482-490.
2. Franke A, Kuhl HP, Rulands D, Jansen C, Erena C, Grabitz RG et al. Quantitative analysis of the morphology of secundum -type atrial septal defects and their dynamic change using transesophageal three-dimensional echocardiography. Circulation 1997; 96:II323-27.
3. Van den Bosch, Harkel DJT, McGhie JS, Roos-Hesselink JW, Simoons ML, Rogers AJJC, Meijboom FJ.

Characterization of Atrial Septal Defect Assessed by Real-time 3-Dimensional Echocardiography. J Am Soc Echocardiogr 2006;19:815-821.

4. Mahmood F, Shakil O, Gerstle JR, Matyal R. Percutaneous Closure of an Atrial Septal Defect and 3-Dimensional Echocardiography. J Cardiothorac Vasc Anesth 2013;27:400-401.
5. Brysiewicz N, Mitiku T, Haleem K, Bhatt P, Al-Shaaroui M, Clancy JR, Marieb MA, Sugeng L, Akar JG. 3D Real-Time Intracardiac Echocardiographic Visualization of Atrial Structures Relevant to Atrial Fibrillation Ablation. J Am Coll Cardiol Img 2014;7:97-100.

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## Letters to the Editor

*Congenital Cardiology Today* welcomes and encourages Letters to the Editor. If you have comments or topics you would like to address, please send an email to: [LTE@CCT.bz](mailto:LTE@CCT.bz), and let us know if you would like your comment published or not.


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# Image of the Month #9: April, 2014 - The Archiving Working Group

Contributors: Jeffrey P. Jacobs, MD; Jorge M. Giroud, MD; Robert Anderson, MD; Vera D. Aiello, MD; Diane E. Spicer, BS; Charles W. Shepard, MD

The Archiving Working Group (AWG) Web Portal link for this series of images: [http://www.accd-awg.umn.edu/Coronary\\_Disease/ALCAPA\\_09\\_41\\_03/ALCAPA\\_09\\_41\\_03\\_MPA.html](http://www.accd-awg.umn.edu/Coronary_Disease/ALCAPA_09_41_03/ALCAPA_09_41_03_MPA.html)

IPCCC: 09.41.03

## AEPC Derived Term:

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) (09.41.03)

## EACTS-STIS Derived Term:

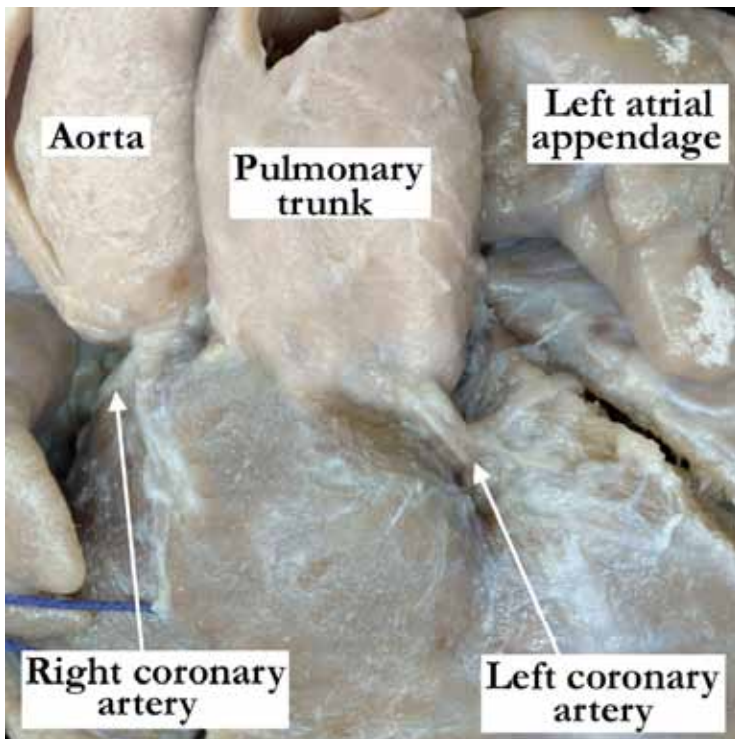
Coronary anomaly, APOC (Anomalous pulmonary origin of coronary), ALCAPA (Anomalous left main coronary artery from PA) (09.41.03)

## ICD10 Derived Term:

Malformation of coronary vessels (Q24.5)

## Commentary

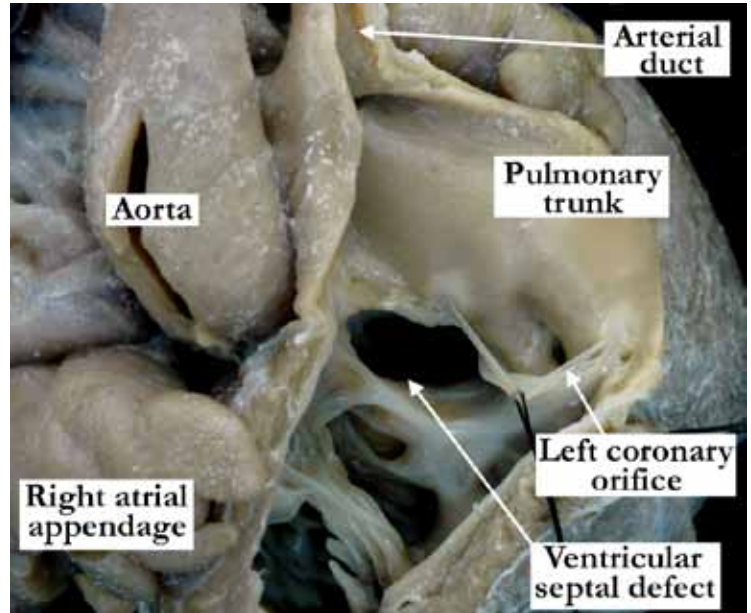
Anomalous pulmonary origin of the left coronary artery is a rare congenital defect involving the coronary circulation. It is estimated to occur in 1 of 300,000 live births. If untreated, it results in



*Orientation:* Anterior superior view

*Description:* This view of a heart with normally related great arteries demonstrates the normal origin of the right coronary artery from the aorta. The left coronary artery arises from the pulmonary right-handed adjacent truncal valvar sinus.

*Contributor:* Diane E. Spicer, BS



*Orientation:* Anterior superior view of the right ventricle, outflow tract and pulmonary trunk

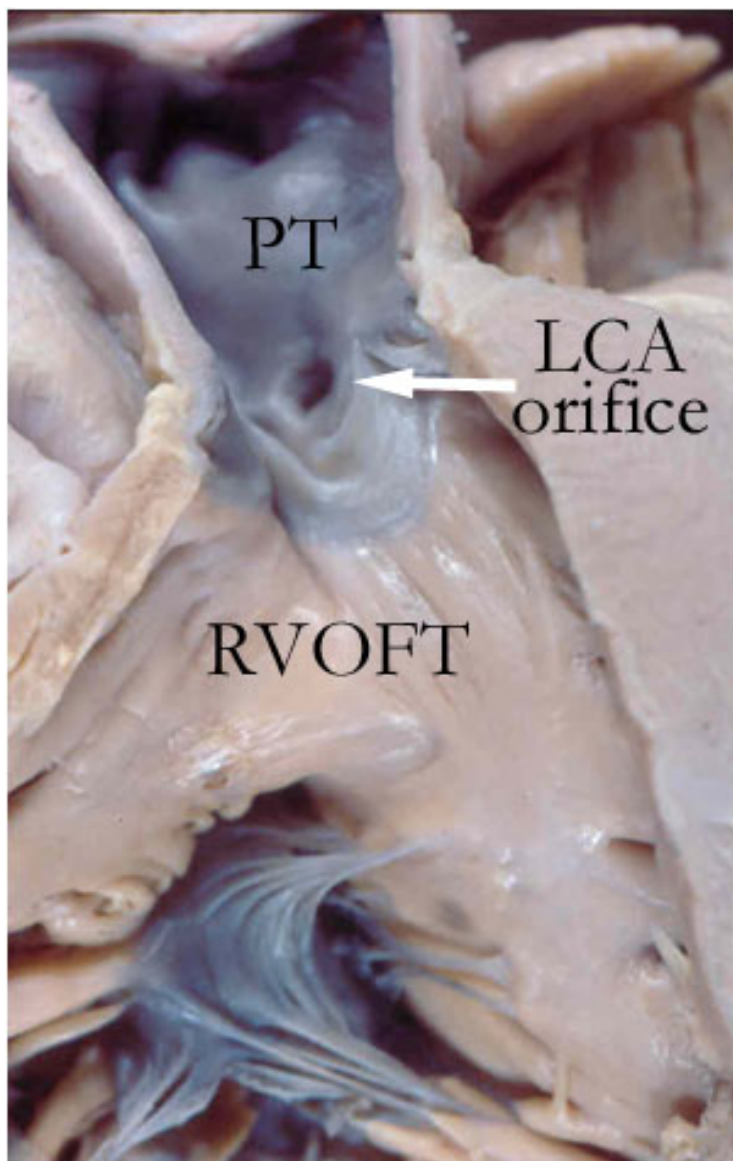
*Description:* This view of the opened right ventricle demonstrates that the left coronary artery arises from the right-handed adjacent sinus of the pulmonary trunk. Well-illustrated in this image is the presence of a doubly committed and juxta-arterial ventricular septal defect. Although not shown in this image, other findings included signs of left ventricular ischemia, severe aortic stenosis secondary to postero-caudal deviation of the outlet septum, interrupted aortic arch distal to the brachiocephalic trunk, and isolated origin of the left carotid artery from the right pulmonary artery, hypoplastic right ventricle and tricuspid valve dysplasia. Please note the patent arterial duct.

*Contributor:* Diane E. Spicer, BS

ischemia, left ventricular dysfunction or infarction accompanied by mitral regurgitation, arrhythmias, and death in infancy in up to 90% of affected patients. The lesion commonly presents in early infancy, and manifests with increasing difficulties in breathing and feeding, accompanied by physical findings of mitral regurgitation and myocardial ischemia. Although there are case reports that date as early as the 19<sup>th</sup> century, it is Bland, White and Garland, from Massachusetts General Hospital, who are credited as being

***“Anomalous pulmonary origin of the left coronary artery is a rare congenital defect involving the coronary circulation. It is estimated to occur in 1 of 300,000 live births. If untreated, it results in ischemia, left ventricular dysfunction or infarction accompanied by mitral regurgitation, arrhythmias, and death in infancy in up to 90% of affected patients.”***





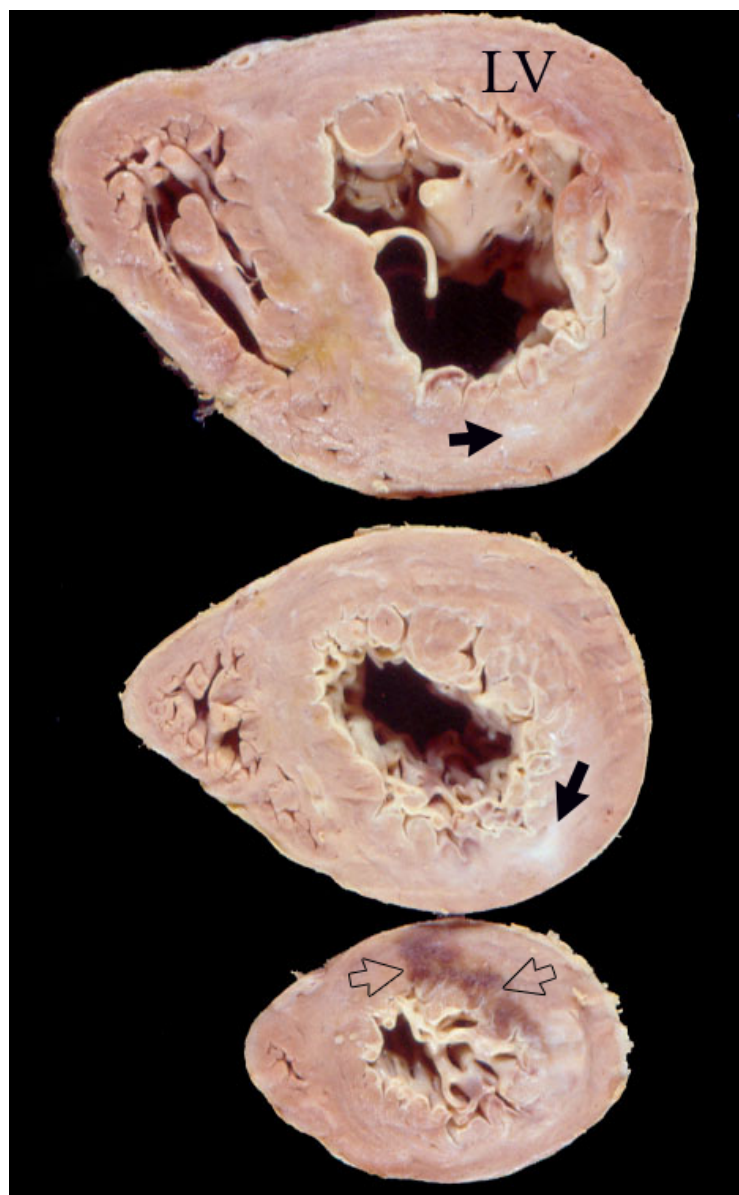
**Orientation:** Anterior-superior view through opened right ventricular outflow tract and pulmonary trunk

**Description:** In a different specimen, the right ventricular outflow tract (RVOFT) has been exposed to show the Anomalous Origin of the Left Coronary Artery (arrow) from the right-handed adjacent sinus of the pulmonary trunk (PT).

**Contributor:** Vera D. Aiello, MD

**Institution & Source:** Heart Institute (InCor), University of São Paulo Medical School, São Paulo, Brazil

the first to describe a constellation of clinical observations, validated by the autopsy findings in their patient. The anatomical observation of an abnormal origin of the left coronary artery from the pulmonary circulation provides understanding of the pathophysiology of the disorder. As the pulmonary vascular resistance falls in early infancy, there are incremental changes in



**Orientation:** Short-axis sections of the ventricular mass

**Description:** In this heart from a child with Anomalous Origin of the Left Coronary Artery from the pulmonary trunk there are signs of chronic and acute ischemic heart disease. The left ventricle (LV) is hypertrophic and the myocardium shows areas of fibrosis (healed infarction, black arrows) besides an apical area of recent necrosis (open arrows).

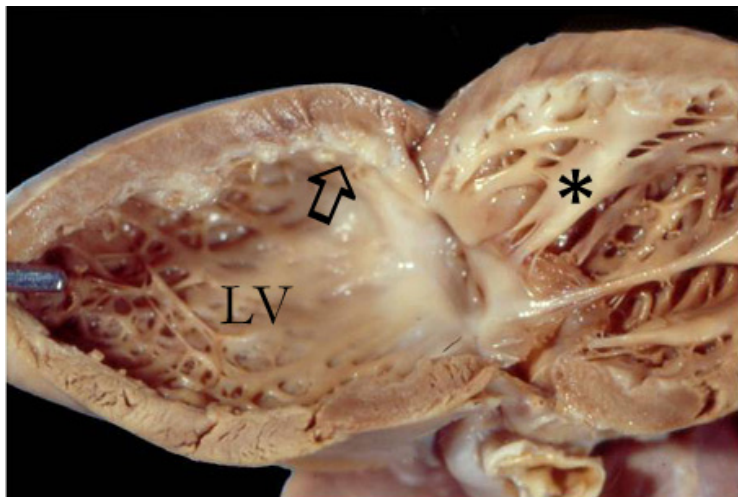
**Contributor:** Vera D. Aiello, MD

**Institution & Source:** Heart Institute (InCor), University of São Paulo Medical School, São Paulo, Brazil

symptoms due to the fall in myocardial perfusion. This 'run-off', or 'steal', of blood away from the ventricle, and into the lesser resistance of the pulmonary circulation, decreases myocardial perfusion. It is thought that, if there is insufficient collateralization from the right coronary circulation, myocardial ischemia with mitral



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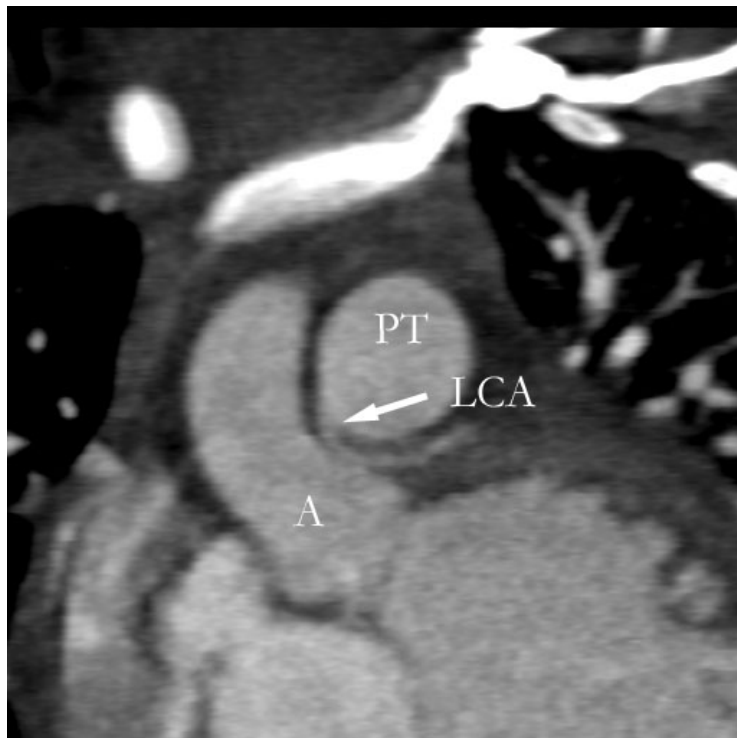
**Orientation:** Left ventricle opened in clamshell fashion.

**Description:** In this heart, from a patient with Anomalous Origin of the Left Coronary Artery, there are signs of long-standing left ventricular (LV) ischemia from the pulmonary circulation. There is severe ventricular dilatation and the subendocardial myocardium shows fibrosis (arrow). The supero-posterior (3) papillary muscle of the mitral valve (asterisk) is pale and fibrotic.

**Contributor:** Vera D. Aiello, MD

**Institution & Source:** Heart Institute (InCor), University of São Paulo Medical School, São Paulo, Brazil

valvar dysfunction increases as time passes, and the pulmonary resistance changes. This can ultimately lead to myocardial infarction, with arrhythmias occurring during periods of increased metabolic demand, such as feeding, crying or an intercurrent illness. Although typically the most common origin of the abnormal left coronary artery is from the pulmonary truncal sinuses, the abnormal origin can also be from the pulmonary trunk, or from the right or left pulmonary arteries. In some instances, the left coronary artery can have a normal origin, but then fail to divide, so that either the circumflex or superior interventricular (anterior descending) artery may arise anomalously from the pulmonary circulation. In the most common form, the abnormal coronary artery arises from a pulmonary valvar sinus, as shown in the initial three images, rather than the pulmonary trunk. The traditional acronym for this entity is ALCAPA or Anomalous Origin of Left Coronary Artery from Pulmonary Artery (or pulmonary trunk). In most cases, however, this is not an accurate representation of the abnormality. Because of these facts, the acronym favored by the surgeons, namely "APOC," or Anomalous Pulmonary Origin of the Coronary Artery, is the more accurate term. This, however, does not distinguish between anomalous origin of the right, as opposed to the left, coronary arteries. APOLC, or Anomalous Pulmonary



**Orientation:** Oblique Coronal CT Angiogram

**Description:** In another patient with Anomalous Pulmonary Origin of the Left Coronary Artery, the coronary artery arises from the pulmonary trunk, approximately 3 mm above the sinutubular junction, and not from a pulmonary valvar sinus. It runs between the trunk and the aorta to reach the interventricular groove. This is an unusual presentation, as the abnormal coronary artery arises from the pulmonary trunk rather than the more common pulmonary valvar sinus origin.

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**Institution:** University of Minnesota Amplatz Children's Hospital

Origin of the Left Coronary Artery, therefore, would be the better acronym. It is doubtful, nonetheless, whether ALCAPA will ever be discarded.

## References

1. J. of Cardiovasc. Trans. Res. (2013) 6:197–207.
2. Pediatr Radiol. 2007 Sep;37(9):890-5.
3. Anderson RH, Loukas M. The importance of attitudinally appropriate description of cardiac anatomy. Clin Anat. 2009 Jan; 22(1):47-51.

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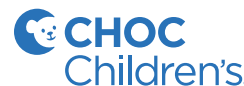
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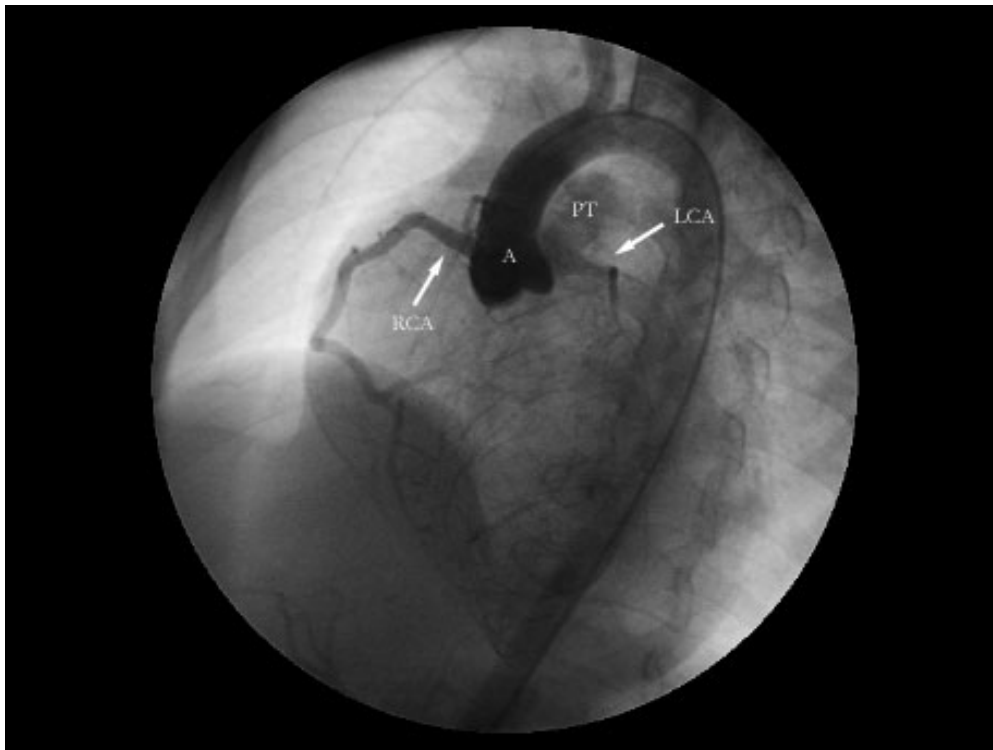
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**Description:** In another patient with Anomalous Pulmonary Origin of the Left Coronary Artery, an aortogram shows that the right coronary artery (RCA) arises from the right coronary aortic sinus and it is larger than usual. The presumed left coronary aortic sinus, in fact, has no coronary artery arising from it. It can be seen that there is opacification of the left coronary artery (LCA) through myocardial collateral channels. The artery arises anomalously from a pulmonary truncal sinus, with the trunk (PT) filled in retrograde fashion.

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# Case Series: Unusual Causes of Dynamic ST-T Segment Changes

By Tabitha G. Moe, MD; Edward K. Rhee, MD; Joseph Graziano, MD

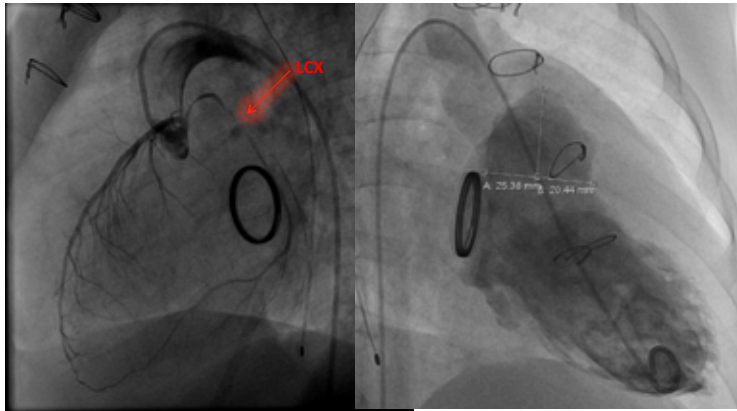


Image 1. Dynamic LCX compression with Diastole. Demonstration of large left atrioventricular disruption with aneurysm.

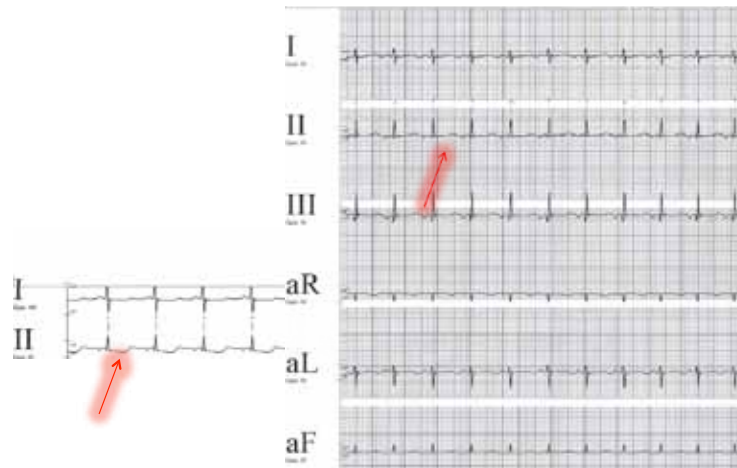


Image 2. Dynamic ST segment depression in lead II.

## Case 1

Case 1 is an 18-month-old female with a history of congenital mitral valve dysplasia with severe mitral regurgitation and mild mitral stenosis who is status post initial palliation at five months of age with a 16 mm St. Jude mitral valve replacement. She developed a thrombus along the valve along in addition to dysfunction of the valve prosthesis and underwent surgical inspection of the mitral valve and prosthetic valve thrombectomy. She developed a post-operative complication of a left middle cerebral artery embolic stroke with complete neurologic recovery. She then developed a left subdural hematoma and underwent craniotomy and hematoma evacuation without significant neurologic deficits. Repeat mitral valve replacement was indicated for redevelopment of mitral valve prosthesis dysfunction and thrombus formation with a 19 mm St. Jude MVR. A follow-up echo showed significant ventricular dysfunction and the patient was referred for cardiac catheterization. Cardiac catheterization showed dynamic diastolic compression of the left circumflex artery by a pseudoaneurysm thought to be an atrioventricular (AV) disruption at the site of her two previously placed prosthetic mitral valves (Image 1). The dynamic diastolic compression causes interesting EKG features consistent with ST-segment elevation

and ischemia (Image 2). Echo analysis revealed progression of wall motion abnormalities, and slowly decrementing left ventricular ejection fraction in the setting of an ongoing coronary insult. Medical and operative management continued to be difficult in light of two failed mitral valve prostheses, and she was subsequently listed for heart transplant. She underwent a bicaval orthotopic heart transplant at age 20 months and was able to be discharged to home without further postoperative complications.

## Case 2

The patient is a 13-year-old male with a history of repaired truncus arteriosus and an interrupted aortic arch status post-arch reconstruction. Cardiac catheterization one year previously noted moderate narrowing of the pulmonary homograft, moderate pulmonary insufficiency, and a peak gradient across his aortic arch of 50 mmHg. He was referred for pulmonary homograft replacement and arch repair. The operative course was unremarkable and he was transferred to the cardiovascular intensive care unit (CVICU) for recovery. Shortly after arrival in the CVICU, he was noted to have 3 mm ST-segment elevation in leads II, III, and aVF suggestive of inferior ischemia (Image 3). The patient's previous cardiac magnetic resonance imaging was reviewed extensively and there were no coronary anomalies noted. It was, therefore, determined that he should be taken to the cardiac catheterization lab to evaluate for patency of the RCA. Fluoroscopic evaluation of coronary anatomy revealed compromise of the RCA by the chest tube placed on intermittent suction for drainage post-operatively (Images 4 and 5). The chest tube was removed and the ST-segment changes resolved.

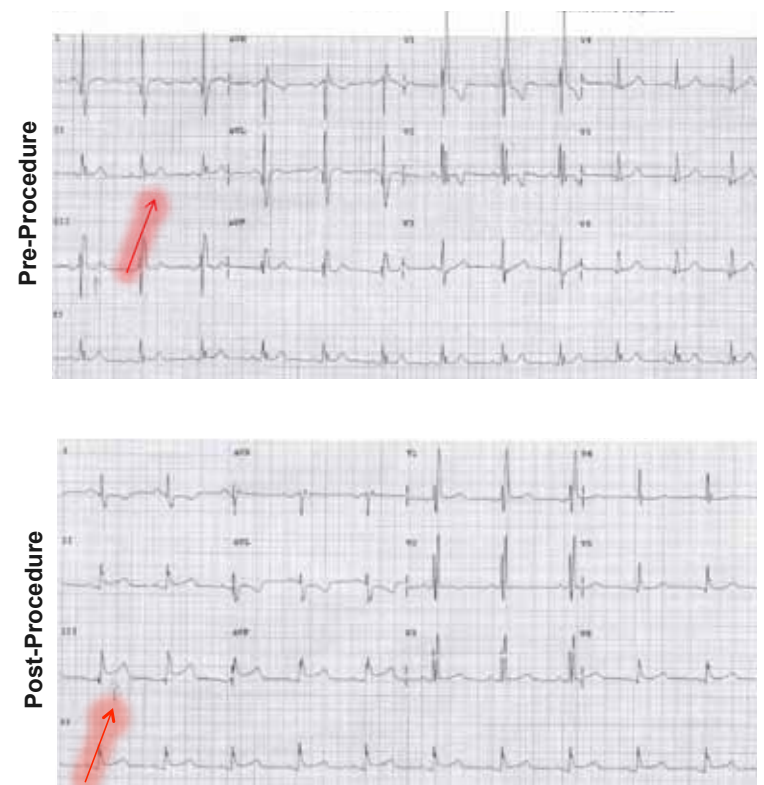


Image 3 - Dynamic LCX compression with Diastole. Demonstration of large left atrioventricular disruption with aneurysm.



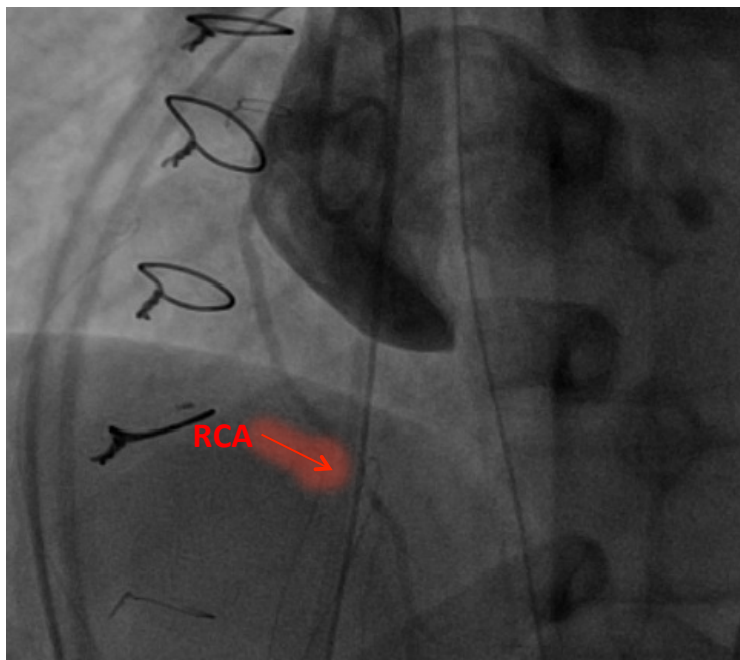


Image 4. Dynamic LCX compression with Diastole. Demonstration of large left atrioventricular disruption with aneurysm.

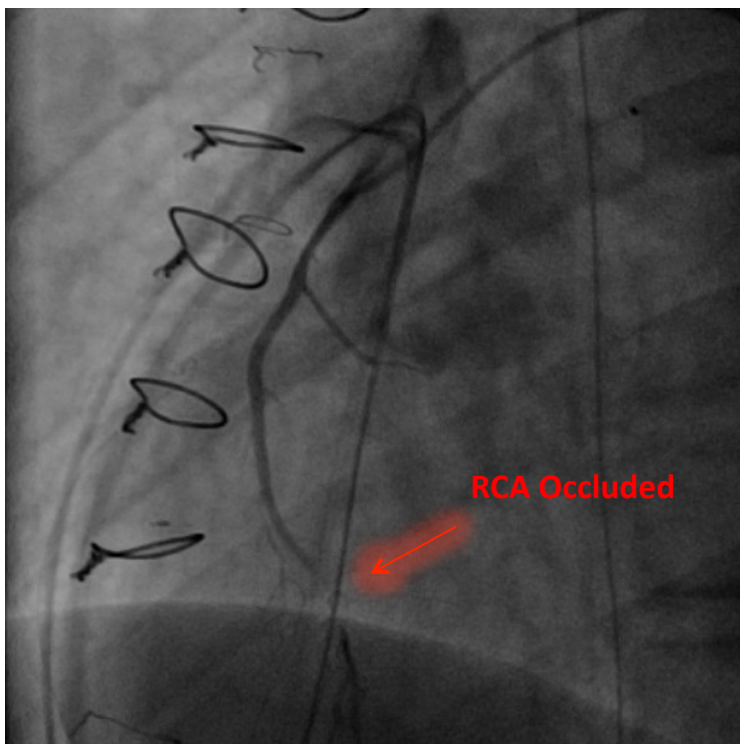


Image 5. Selective RCA angiography with mid RCA occlusion.

## Discussion

These two unique cases highlight the need for clinical acumen in electrocardiogram interpretation. Each of these cases presents a very unusual set of circumstances leading to intermittent myocardial ischemia in a child. Atrioventricular (AV) disruption occurs rarely (1% to 2%), but is usually fatal with an operative mortality rate of 50%.<sup>1,2</sup> Our patient is unique in that her presentation of AV disruption was found during cardiac catheterization after an abnormal echo suggested it. In the case of multiple mitral valve surgeries, AV disruption, injury to the left circumflex artery and thromboembolic events are infrequent but documented complications.<sup>3</sup> Various predisposing and intraoperative factors have been suggested as to the cause of AV disruption.<sup>4</sup> Dark and Bain suggest the cause of the rupture at the AV groove occurs after damage to the myocardium that has lost the internal support structure of the subvalvular apparatus.<sup>5</sup> Although there are multiple case reports of left coronary artery injury or accidental occlusion perioperatively, there are no documented cases of dynamic left circumflex occlusion secondary to AV disruption.<sup>6-10</sup> There are rare case reports of patients developing acute ischemia following uneventful non-coronary cardiac surgery even after successful weaning of cardio-pulmonary bypass in adults. There are exactly two cases of coronary ischemia reported secondary to chest tube placement with intermittent suction.<sup>11</sup> A similar case reports RCA acute marginal branch compression by intrapericardial drain placement and not an extra-cardiac chest tube with intermittent suction.<sup>12</sup> There are no previously reported cases of RCA compression in children after conduit replacement. These two cases serve as excellent reminders to critically examine both the patient and objective data in patients with complex congenital heart disease, and multiple repairs.

The authors have no disclosures.

***“There are no previously reported cases of RCA compression in children after conduit replacement. These two cases serve as excellent reminders to critically examine both the patient and objective data in patients with complex congenital heart disease, and multiple repairs.”***

## References

1. Karlson KJ, Ashraf MM, Berger RL. Rupture of left ventricle following mitral valve replacement. *Ann Thorac Surg* 1988;46: 590–7.
2. Bjork VO, Henze A, Rodriguez L. Left ventricular rupture as a complication of mitral valve replacement. *J Thorac Cardiovasc Surg* 1977;73:14–22.
3. Cammack PL, Edie RN, Edmunds LH Jr. Bar calcification of the mitral annulus: a risk factor in mitral valve operations. *J Thorac Cardiovasc Surg* 1987;94:399–404.



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4. Katske G, Golding LR, Tubbs RR, Loop FD. Posterior midventricular rupture after mitral valve replacement. *Ann Thorac Surg* 1979;27:130-2.
5. Dark JH, Bain WH. Rupture of posterior wall of left ventricle after mitral valve replacement. *Thorax* 1984;39:905-11.
6. Morin D, Fischer AP, Sohl BE, Sadeghi H. Iatrogenic myocardial infarction. A possible complication of mitral valve surgery related to anatomical variation of the circumflex coronary artery. *Thorac Cardiovasc Surg* 1982;30:176-9.
7. Kaklikkaya I, Yeginoglu G. Damage to coronary arteries during mitral valve surgery. *Heart Surg Forum* 2003;6:E138-42.
8. Cornu E, Lacroix PH, Christides C, Laskar M. Coronary artery damage during mitral valve replacement. *J Cardiovasc Surg (Torino)* 1995;36:261-4.
9. Virmani R, Chun PK, Parker J, McAllister HA Jr. Suture obliteration of the circumflex coronary artery in three patients undergoing mitral valve operation. Role of left dominant or codominant coronary artery. *J Thorac Cardiovasc Surg* 1982;84:773-8.
10. Danielson GK, Cooper E, Tweedale DN. Circumflex coronary artery injury during mitral valve replacement. *Ann Thorac Surg* 1967;4:53-9.
11. Chapin JW, Kahre J, Newland M. Acute Myocardial Ischemia caused by mediastinal chest tube suction. *Anesth Analg* 1980; 59:386-7.
12. Sulimovic S, Noyez L. Postoperative myocardial ischemia caused by compression of a coronary artery by a chest tube. *J Cardiovasc Surg* 2006; 47:371-2.

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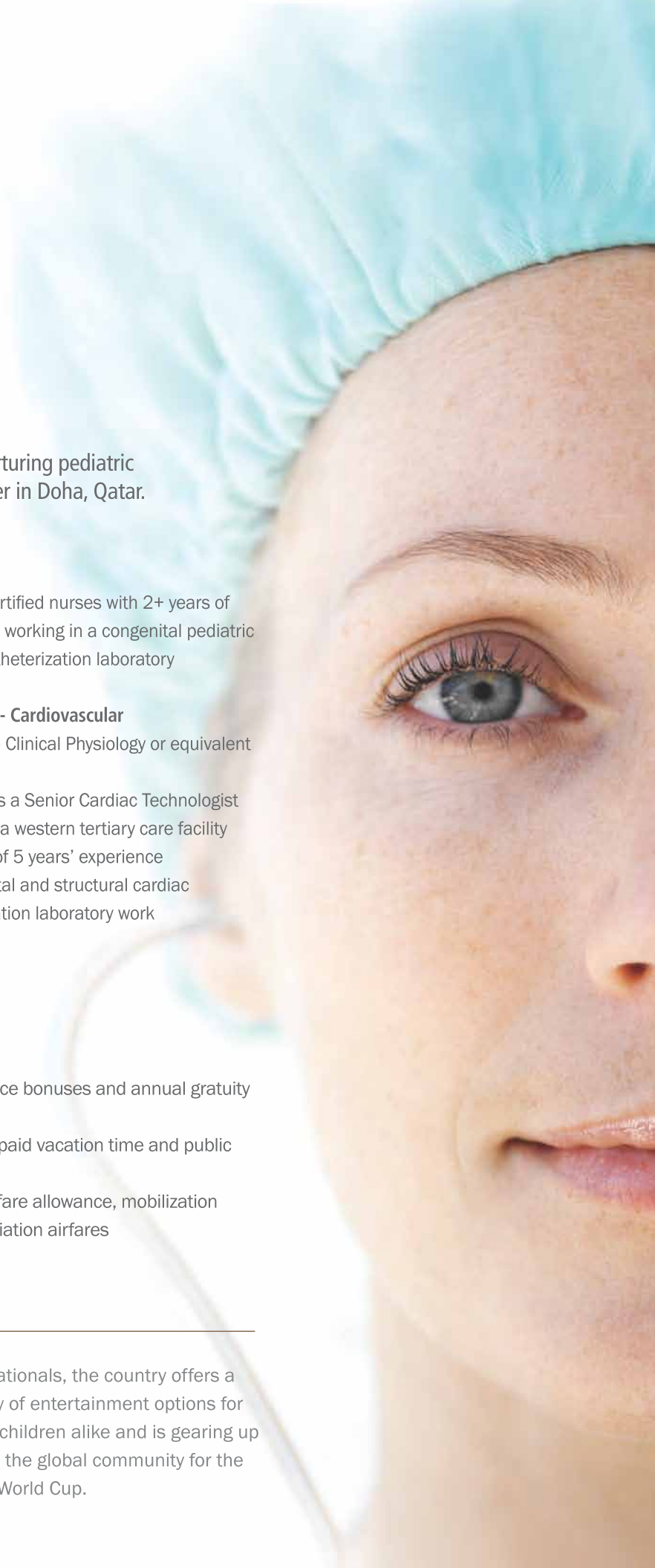
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