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. 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 electrophysiological procedures. 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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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.

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.

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.
JUNE MEDICAL MEETING FOCUS
Fetal Cardiac Symposium
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A 2-day basic and advanced fetal cardiology symposium and workshop offering thorough and updated presentations on scanning the fetal heart and diagnosing and managing various common fetal congenital heart disease malformations. It will emphasize the basics of fetal cardiac scanning coupled with live case demonstrations and tips for diagnosing various anomalies. Presentations will focus on anomalies of the four-chamber and outflow-tracts views, reflecting the recent guidelines for screening for fetal heart disease.

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-td 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.
The Melody® TPV offers children and adults a revolutionary option for managing valve conduit failure without open heart surgery. Just one more way Medtronic is committed to providing innovative therapies for the lifetime management of patients with congenital heart disease.

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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 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 imaging in the assessment of patients during procedures for interventional device closure. Figure 5C 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.
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 22nd Annual International Echo Symposium in Parma, Italy, June 16th-18th, 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


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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.biz, and let us know if you would like your comment published or not.
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

IPCCC: 09.41.03

AEPC Derived Term: Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) (09.41.03)

EACTS-STS 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 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 19th century, it is Bland, White and Garland, from Massachusetts General Hospital, who are credited as being the first to describe this condition.

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

“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.”
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 trunk provides understanding of the pathophysiology of the disorder. As the pulmonary vascular resistance falls in early infancy, there are incremental changes in 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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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

Please visit us at the AWG Web Portal at http://ipccc-awg.net/ and help in the efforts of the Archiving Working Group and the International Society for Nomenclature of Paediatric and Congenital Heart Disease.

The authors would like to acknowledge the Children’s Heart Foundation (http://www.childrensheartfoundation.org/) for financial support of the AWG Web Portal.
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

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

Image 2. Dynamic ST segment depression in lead II.

Image 3 - Dynamic LCX compression with Diastole. Demonstration of large left atrioventricular disruption with aneurysm.
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%.1,2 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.3 Various predisposing and intraoperative factors have been suggested as to the cause of AV disruption.4 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.5 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.6-10 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.11 A similar case reports RCA acute marginal branch compression by intrapericardial drain placement and not an extra-cardiac chest tube with intermittent suction.12 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


Image 5. Selective RCA angiography with mid RCA occlusion.
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