

Coronary Artery Ostial Atresia

Diagnosis of Conotruncal Anastomotic Collateral Rings Using CT Angiography

Farhood Saremi, MD,* Graydon Goodman, MS,* Alison Wilcox, MD,* Raffi Salibian, MD,*
Gabriel Vorobiof, MD†‡

IN THIS PAPER WE DISCUSS 2 UNPRECEDENTED EXAMPLES OF CORONARY OSTIAL ATRESIA IN ADULTS. The first case documents a case of left main ostial agenesis with intercoronary communication through a retroaortic Kugel anastomotic collateral. The second case describes a right coronary artery (RCA) ostial agenesis with a preconal intercoronary anastomosis in association with supraaortic pulmonary stenosis. While atresia of the left coronary artery ostium is a rare anatomic variant of the coronary circulation, atresia of the right coronary ostium is exceedingly rare and not reported in adults (1). Correlative images of coronary computed tomography angiography (CTA) using a 64-slice multidetector computed tomography and coronary catheterization angiograms are presented for both cases, and the arterial collateral pathways between the proximal right and left coronary systems are discussed. We also reviewed the differences between congenital and developmental forms of abnormalities.

Atresia of the left coronary ostium. A 67-year-old Hispanic woman who was a nonsmoker without significant past medical history was admitted for the evaluation of 3 month history of exertional angina and dyspnea. The patient reported worsening of symptoms when walking farther than 1 block or heavy lifting. She denied resting chest pain. Her resting electrocardiogram (ECG) was normal, however an office stress echocardiogram performed prior to admission was terminated after 3 min due to chest pain, shortness of breath, and 1-mm ST-segment depression. The post-exercise echocardiogram showed mild global left ventricle hypokinesis and no increase in left ventricular size. Cardiac catheterization was performed. During initial aortogram the left main coronary artery was not opacified, even after direct injection of contrast medium into the left sinus of Valsalva. However, a large collateral artery originating from the ostium of the RCA was identified on the RCA injection and subsequent selective catheterization of its orifice (Fig. 1). This 3-mm diameter collateral vessel continued posteriorly behind the aortic root and connected to the proximal portion of the left circumflex (LCX) artery and supplied the entire left coronary arterial tree in an antegrade fashion. A smaller collateral ring between the right and left conus arteries was also shown connecting the proximal RCA to the left anterior descending (LAD). No connection between the left main and the left coronary cusp was observed, but instead, the ostium of the left main artery formed a blind loop. The RCA had an enlarged caliber but the LAD and the LCX were normal in size. Mild left coronary artery luminal irregularities were observed.

From the *Department of Radiology, University of Southern California, Los Angeles, California; †Heart and Vascular Institute, Memorial Care Medical Center, Long Beach, California; and the ‡Division of Cardiology, University of California Irvine, Orange, California. Dr. Vorobiof has served on the Speakers' Bureau for Lantheus Medical Imaging. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

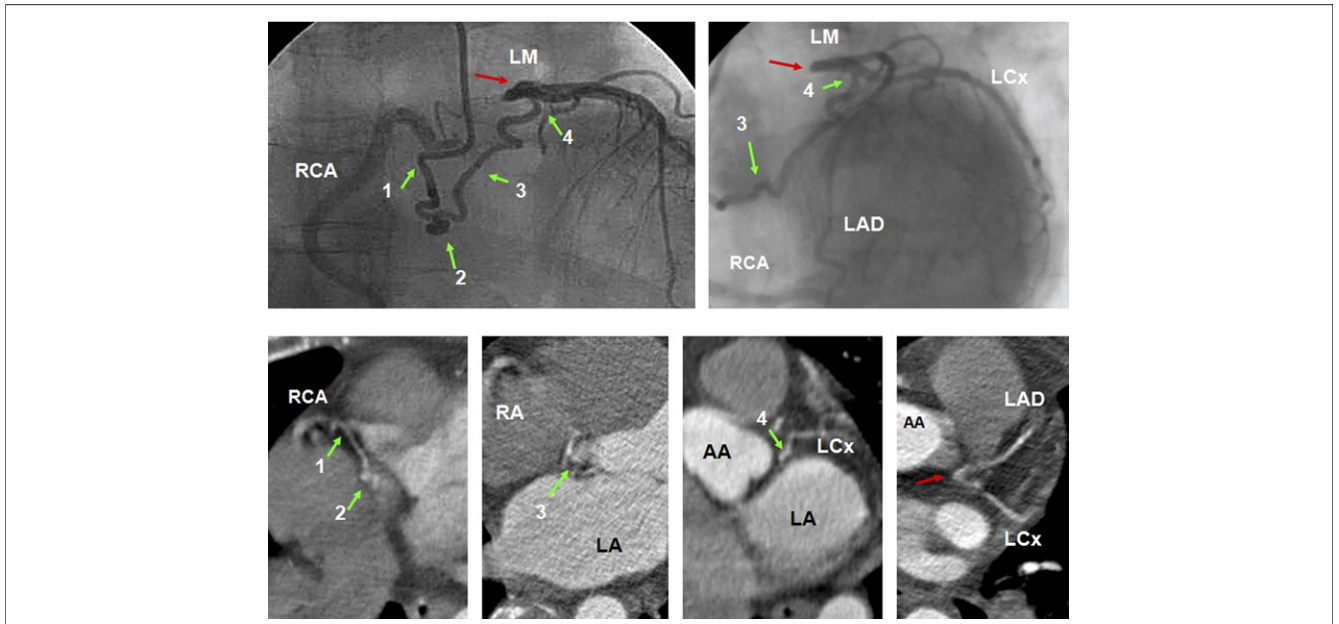


Figure 1. Left Coronary Ostium Atresia

Left main (LM) atresia with retroaortic intercoronary collateral pathway provided by Kugel anastomotic ring. The **top row** shows coronary angiograms with selective injection into the collateral artery origination near the ostium of the right coronary artery (RCA). The **bottom row** shows axial computed tomography angiography images at different levels showing segments of the anastomotic Kugel artery (**green arrows**). The anastomotic artery is tortuous and difficult to show in entirety in one computed tomography image. The artery starts at the ostium of the RCA and dives posteriorly along the right lateral and then behind the aortic root (**segments 1 to 3**) before travelling in the transverse sinus to connect to the proximal left circumflex (LCx) artery (**segment 4**). **Segment 3** of the Kugel artery is retroaortic and very close to the interatrial septum where it may send branches to the atrioventricular node artery (not seen in this case). The LM is atretic and forms a blind end (**red arrows**) located 5 mm from the left coronary sinus. All other vessels are patent. The left anterior descending (LAD) artery showed mild atherosclerotic disease. The RCA was ectatic. AA = ascending aorta; LA = left atrium; RA = right atrium.

Due to these findings, coronary CTA was requested to further evaluate the coronary catheterization observations and assess for associated structural abnormalities. Coronary computed tomography was performed using a 64-slice scanner and standard retrospective ECG gating. The aortic sinuses were normal in orientation. The left coronary ostium was not seen arising from the left aortic sinus, however, a normal caliber blind-ended left main coronary artery was seen branching into normal caliber LCX and LAD arteries. The LAD demonstrated noncalcified plaque with associated mild stenosis. The blind wall of the left main artery was located 5 mm from the aortic wall. The RCA was mildly dilated. A collateral artery originated from the first centimeter of the RCA, coursed posteriorly to the aortic root, and curved leftwards in the transverse sinus to supply the proximal LCX artery. The anatomic course of this collateral vessel was consistent with an intercoronary anastomotic network of Kugel connecting the proximal RCA to LCX artery.

Atresia of the right coronary ostium. A 46-year-old Hispanic man who was a nonsmoker and had a vague history of prior heart surgery for pulmonary valve disease was admitted for palpitations and lightheadedness. Upon presentation, the patient was found to have ventricular tachycardia at a rate of 186 beats/min and was successfully cardioverted to a normal sinus rhythm of 60 beats/min. The patient was ruled out for myocardial infarction and admitted to the coronary care unit for monitoring and further work up. Right heart catheterization showed a right ventricle with normal morphology, thickened trabeculations, and an estimated ejection fraction of 40%. Mild pulmonary valve stenosis with a peak pressure gradient of 23 mm Hg, moderate post-stenotic dilatation of the main pulmonary artery, and moderate tricuspid regurgitation were also noted. Pulmonary pressures and vascular resistances were normal. Coronary catheterization showed diffuse ectasia of the coronary

arteries (up to 10 mm LAD and 8 mm RCA) with no atherosclerotic disease and the RCA arising from the proximal LAD (Fig. 2).

Coronary CTA was performed to further investigate the coronary anatomy using a 64-slice scanner and standard retrospective ECG gating protocol. The origin of the RCA was not identified arising from the right aortic cusp, instead it was seen as a blind-ended vessel approximately 5 mm from the aorta and that immediately bifurcated into a large trunk of the main RCA, and a large right conus artery measuring 4 mm in diameter (Fig. 2). The RCA then continued its normal anatomic course as a dominant branch. An enlarged collateral conus branch traveled anterior to the main pulmonary artery and joined the proximal LAD (forming Vieussens' ring). All coronary arteries were ectatic, with the LAD measuring approximately 8-mm in diameter without any significant atherosclerosis. The right ventricle, including the outflow tract, was hypertrophied. There was moderate enlargement of the right atrium. The pulmonary arteries appeared patent, however there was 30% annular supravalvular narrowing of the main pulmonary artery approximately 12 mm distal to the pulmonary valve, which measured 14 × 12 mm. The pulmonary valve annulus measured 17 × 18 mm. There was evidence of pulmonary regurgitation and the regurgitant area was 4.6 mm². The patient was stabilized, placed on Amiodarone, and scheduled for follow-up in cardiology clinic.

Discussion

Three major collateral pathways at the conotruncal level provide circulation between the right and left coronary systems in all congenital or acquired forms of one sided coronary artery occlusion and are used as the basis for different classifications (Fig. 3). The right anterior conus artery is the most constant and conspicuous branch participating in the preconal collaterals also known as Vieussens' arterial ring. The Kugel anastomotic artery is an uncommon but sometimes important retroaortic collateral between

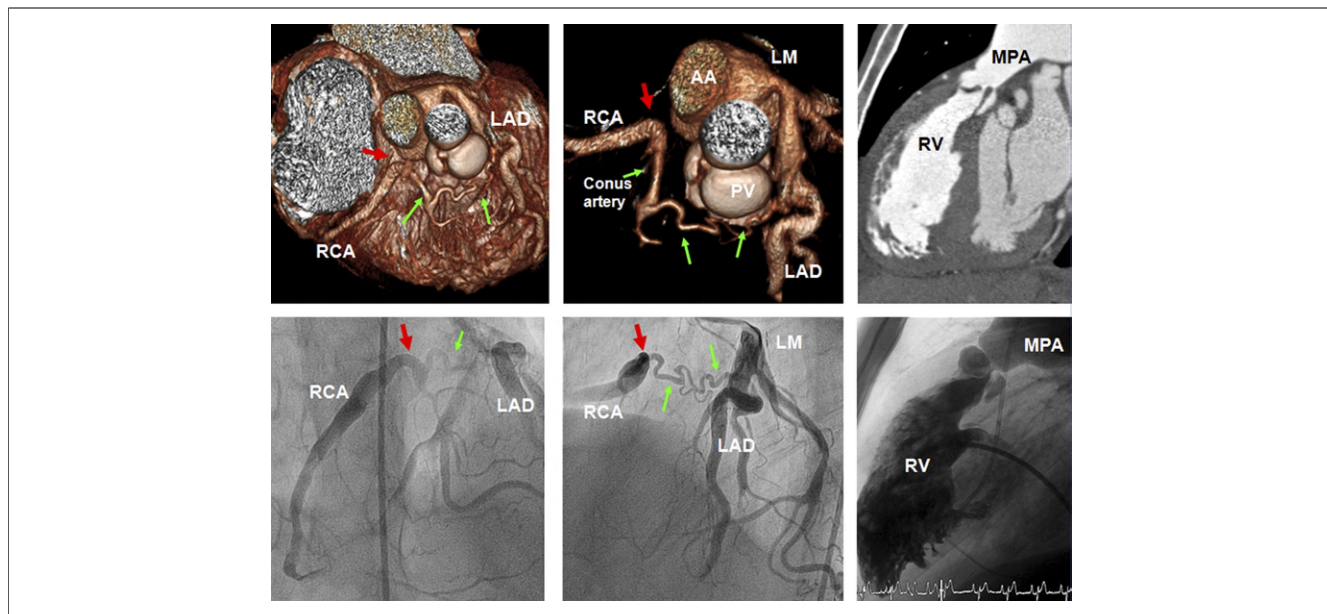


Figure 2. Right Coronary Ostium Atresia

RCA ostial atresia and supravalvular stenosis of the main pulmonary artery (MPA). Computed tomography images are demonstrated (top row) and corresponding coronary catheterization angiography (injection is into the left main artery) and right ventriculogram images are shown (bottom row). A large right anterior conus artery arising from the proximal RCA forms the Vieussens' ring (green arrows) which communicates with the proximal LAD. Note atresia of the RCA at the origin with a blind-ended vessel located 5 mm from the aortic root (red arrows). Both RCA and LAD are ectatic. The right ventricle (RV) is hypertrophied, the pulmonary valve (PV) leaflets are thickened, and mild supravalvular stenosis is seen. Abbreviations as in Figure 1.

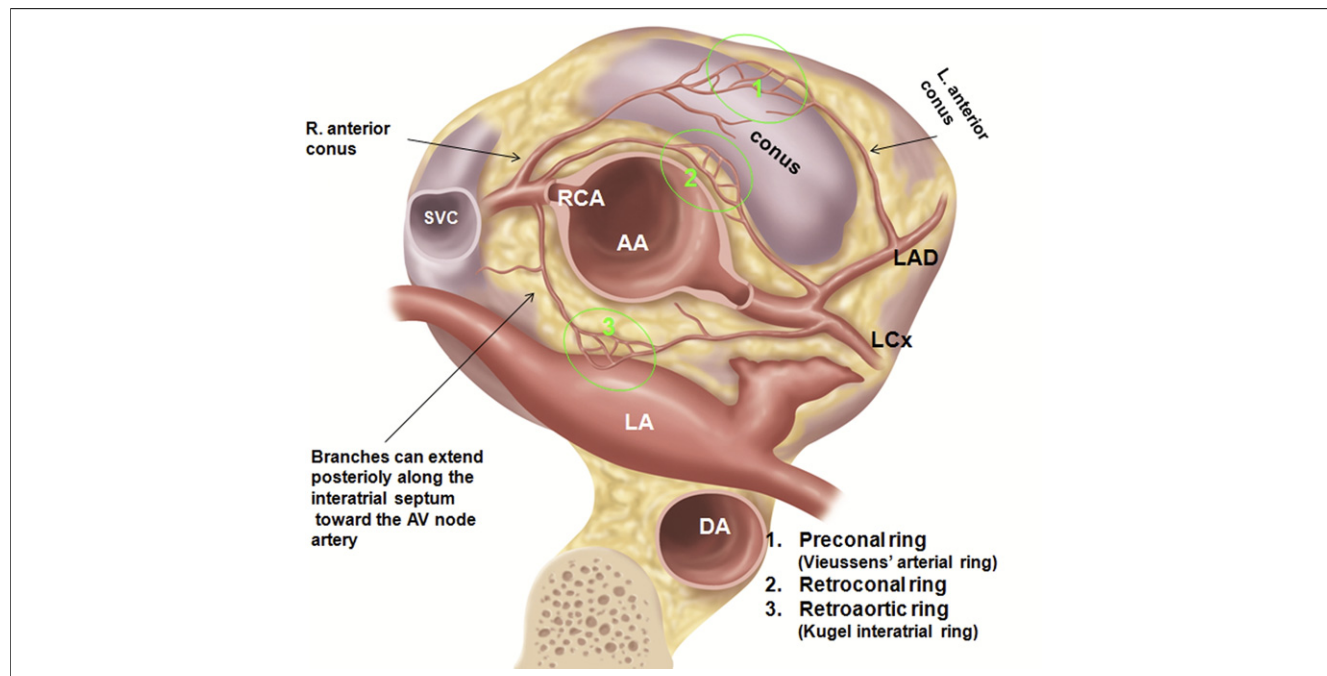


Figure 3. Conotruncal Arterial Anastomotic Circulation

Schematic drawing demonstrates 3 arterial anastomotic rings participating in vascular supply to the aortopulmonary trunk as well as collateral circulation between the proximal right and left coronary artery systems. The preconal ring (circle #1), also known as Viessens' arterial ring, comprises right pulmonary conus branches arising from the RCA or the aorta and left conus branches arising from the left main or proximal LAD arteries. The right anterior conal branch exists in 100% of cases and in 50% it may arise from the right aortic sinus. Left anterior conus exists in 85% of cases, right posterior in 15%, and left posterior in 15%. The retroconal anastomotic ring (circle #2) forms connections from the right and left coronaries through collaterals which traverse in the interarterial space behind the MPA but anterior to the ascending aorta. The retroaortic anastomotic ring, otherwise known as the Kugel atrial anastomotic network, communicates the proximal right and left coronary systems together (circle #3) or individually with a distal coronary system through the interatrial septum (not shown). AV = atrioventricular; DA = descending aorta; SVC = superior vena cava; other abbreviations as in Figures 1 and 2.

the proximal LCX and/or RCA, and the distal coronaries including the atrioventricular node artery via the interatrial septum. Distinguishing ostial atresia from a single coronary artery is important since the single coronary artery is a benign asymptomatic anomaly. Single coronary artery cases are most likely congenital due to early embryonic misconnection of the coronary arteries to the aorta while atresia of the coronary ostium probably occurs at a later stage of life (2).

Address for Correspondence: Dr. Farhood Saremi, Department of Radiology, University of Southern California University Hospital, Keck School of Medicine, 1500 San Pablo Street, Los Angeles, California 90033. *E-mail:* fsaremi@usc.edu.

REFERENCES

1. Musiana A, Cernigliarob C, Sansab M, Masellia D, De Gasperisa C. Left main coronary artery atresia: literature review and therapeutical considerations. *Eur J Cardiothorac Surg* 1997; 11:505-4.
 2. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. *Radiology* 1979;130: 39-47.