Single Coronary Artery: An Unwonted Incidental in a 54-year-old Chronic Kidney Disease Filipino Female with ST Elevation Myocardial Infarction in Cardiogenic Shock

Renato C. Ong, Jr., *RMT, M.D., FPCP, DPCC*,[1] Charles Tadeo O. Galang, *MD, FPCP, FPCC*[1]
[1] Department of Medicine, Section of Cardiology, Makati Medical Center, Philippines
Main Author: Renato C. Ong, Jr.
Contact Information: r.ongjrmd@gmail.com

ABSTRACT

INTRODUCTION
A single coronary artery is extremely rare. Patients with this coronary anomaly are usually asymptomatic and is commonly an incidental finding among imaging diagnostics.

CASE REPORT
We report a case of 56-year-old hypertensive, diabetic Filipino female on thrice-weekly maintenance hemodialysis who had a single origin coronary artery on invasive coronary angiography after she developed STEMI in cardiogenic shock along the course of her admission for an elective below knee amputation. Advised revascularization via coronary artery bypass grafting, however refused. She was eventually discharged against medical advice upon stabilization. She refused further renal replacement therapy; she succumbed two weeks later.

LEARNINGS
Single coronary artery is a rare condition associated with increased risk of sudden cardiac death. A multidisciplinary team in place is ideal to determine the best management strategy to employ. Guideline-directed medical therapy with aggressive risk factor control offers benefit.

KEYWORDS
Single coronary artery. SCA. Cardiogenic shock. ST-elevation myocardial infarction. STEMI.

INTRODUCTION

A rare congenital coronary anomaly that is frequently found incidentally, single coronary artery (SCA) has an incidence of <0.07% among patients undergoing invasive coronary angiography.[1-4]

CASE REPORT

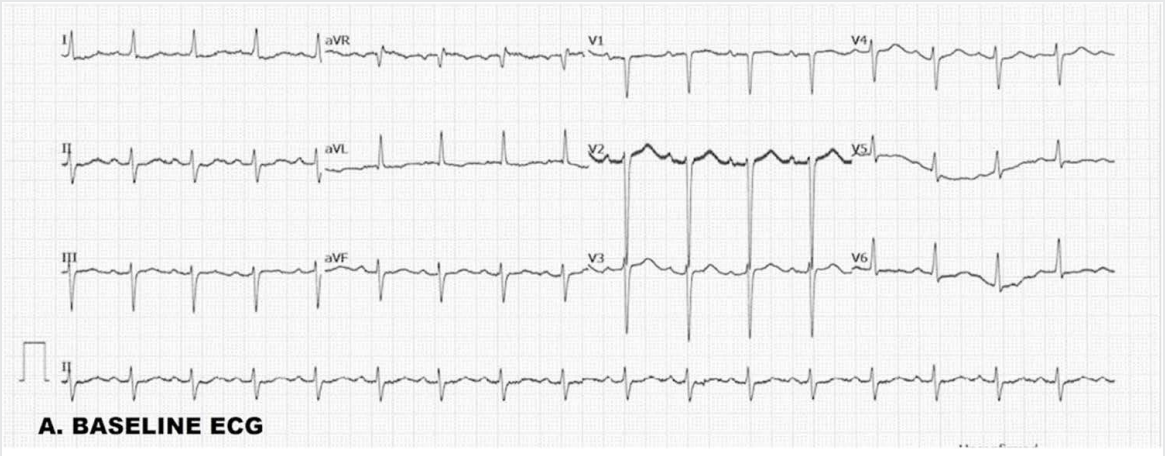
A 54-year-old Filipino female with hypertension, diabetes, bilateral lower extremity arterial occlusive disease, and on maintenance thrice-weekly renal replacement therapy came in for gangrenous right foot.

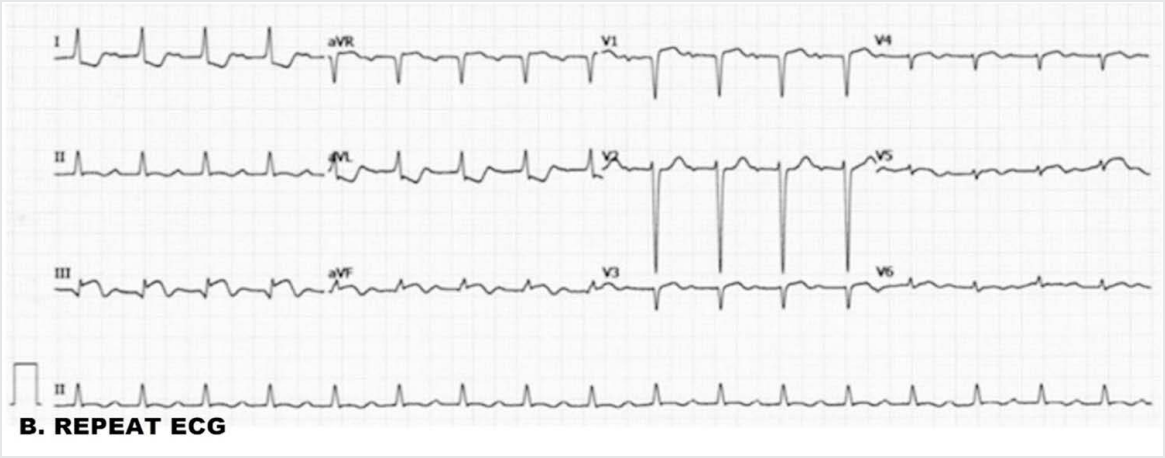
She was worked up at the outpatient department prior to an elective below knee amputation (BKA) after being treated with broad spectrum antimicrobials. Baseline transthoracic echocardiogram (TTE) was concentric left ventricular remodeling with normal wall motion and contractility and a normal left ventricular ejection fraction (LVEF), with mild to moderate mitral and tricuspid regurgitation. Electrocardiogram (ECG) showing sinus tachycardia at 102 bpm, left axis deviation, left ventricular hypertrophy by voltage, abnormal R wave progression, prolonged QTc, nonspecific ST-T wave changes (Figure 1A). She was stratified as having intermediate risk probability, she eventually underwent an unremarkable procedure.

However, a day prior to discharge, she developed sudden hypotension to as low as 70/50 mmHg with no other associated symptoms (i.e, chest pain, palpitations, dyspnea, dizziness) nor other unusual findings. She necessitated norepinephrine infusion after refractoriness to fluid resuscitation. Repeat ECG showed sinus tachycardia (103 bpm), first degree AV block (PR interval 0.28 msec), new onset 1-3 mm ST elevation in leads II, III, aVF with reciprocal 2-3 mm rapid downsloping ST depression in leads I and aVL, prolonged QTcB (0.47 msec) and nonspecific ST-T wave changes (Figure 1B). Repeat TTE showed eccentric left ventricular hypertrophy with moderate global hypokinesis, a mildly reduced LVEF (47%), moderate mitral and tricuspid regurgitation, mild pulmonary hypertension and grade III left ventricular diastolic dysfunction with elevated filling pressures. Cardiac biomarkers were all significantly increased: 1,507x high sensitivity Troponin I, 1.28x total creatine kinase and 3.2x CK-MB.

She was given low molecular weight heparin and dual antiplatelet therapy, and underwent percutaneous coronary angiography, showing a very short single origin of the coronary arteries which immediately trifurcates to a 2.5 mm calcified left anterior descending artery (LAD) with 40% ostial stenosis with myocardial bridging; a 1.75 mm left circumflex artery (LCX) with 70-80% ostial narrowing; and a 5 mm heavily calcified right coronary artery (RCA) with 99% ostial stenosis (Figure 2). The Syntax score of 33+ was computed [coronary artery bypass grafting (CABG), 26.8% vs. percutaneous coronary intervention (PCI), 44%; $p<0.001$], she was advised surgical revascularization through coronary artery bypass grafting, however refused. After the procedure, a multidisciplinary team meeting discussing the best options for the patient, but she opted to be discharged against medical advice. ECG prior to discharge showed sinus rhythm, abnormal R wave progression, first degree AV block, high lateral and lateral wall ischemia, nonspecific ST-T wave changes (Figure 1C). After tapering off norepinephrine, the patient's decision was respected and was sent home with guideline-directed medical therapy with advice of close follow up and aggressive control of risk factors. She also refused further outpatient hemodialysis treatment; two weeks after discharge, she succumbed.

Figure 1: Electrocardiograms. A. Baseline ECG. sinus tachycardia at 102 bpm, left axis deviation, left ventricular hypertrophy by voltage, abnormal R wave progression, prolonged QTc, nonspecific ST-T wave changes. B. Repeat ECG. It showed sinus tachycardia at 103 bpm, first degree AV block at PR 0.28 msec, a new onset 1-3 mm ST elevation in leads II, III, aVF with reciprocal 2-3 mm rapid downsloping ST depression in leads I and aVL, a prolonged QTcB at 0.47 msec and nonspecific ST-T wave changes. C. ECG prior to discharge. Sinus rhythm, abnormal R wave progression, first degree AV block, high lateral and lateral wall ischemia, nonspecific ST-T wave changes.





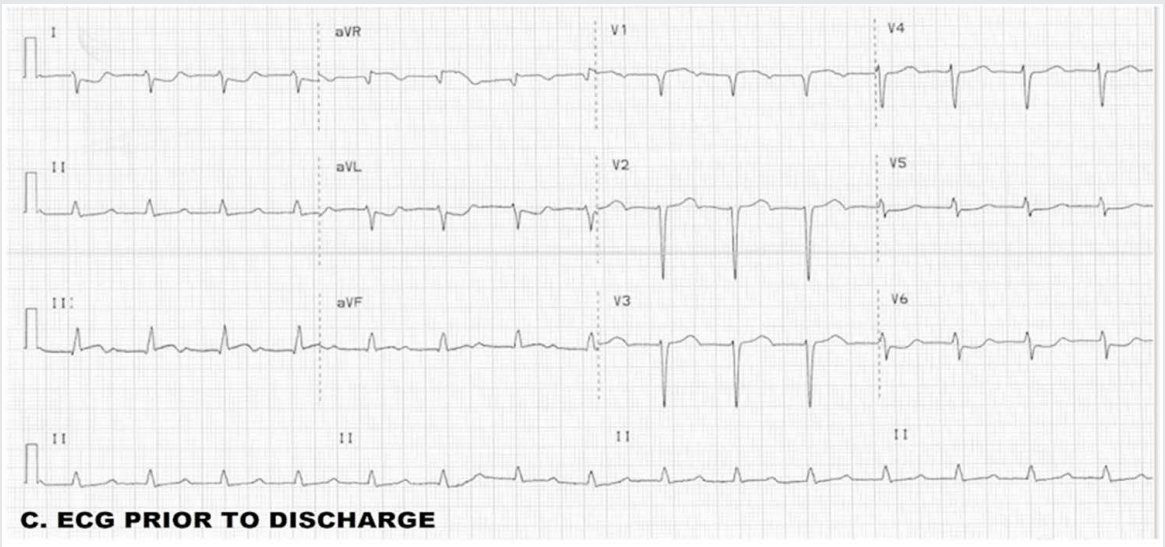
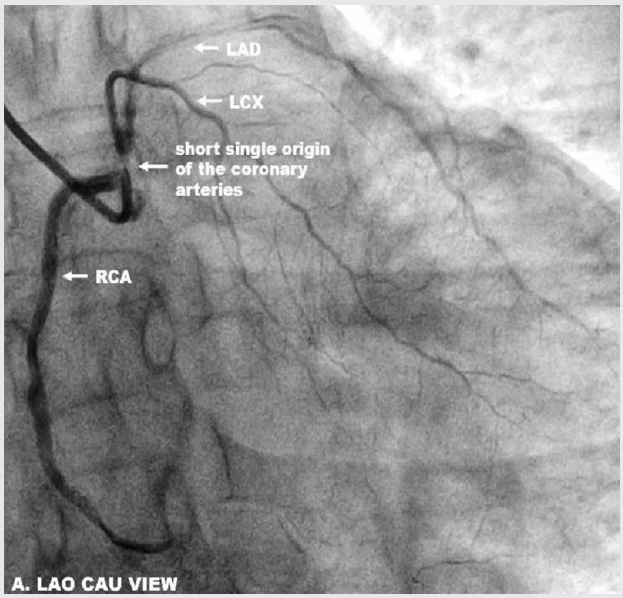
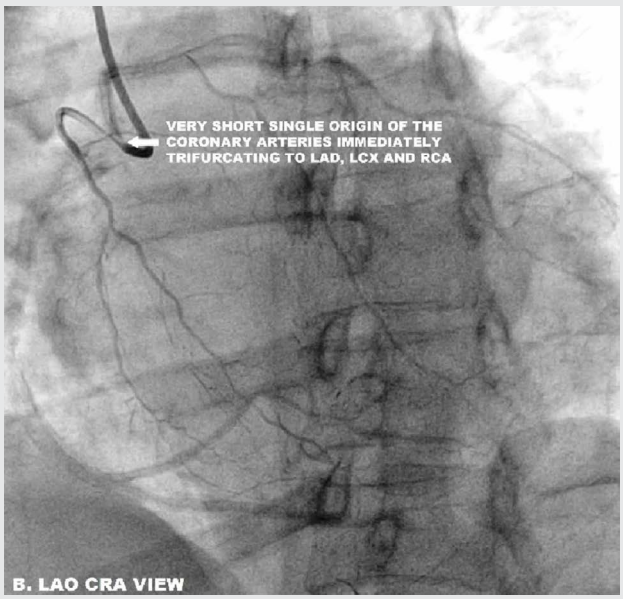
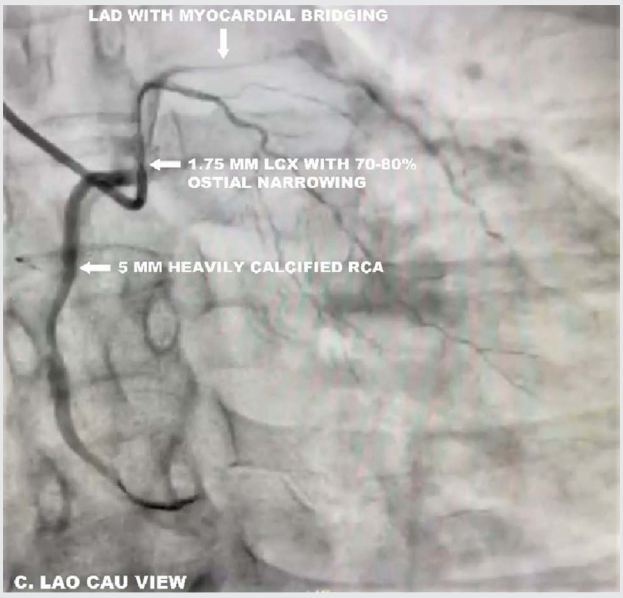


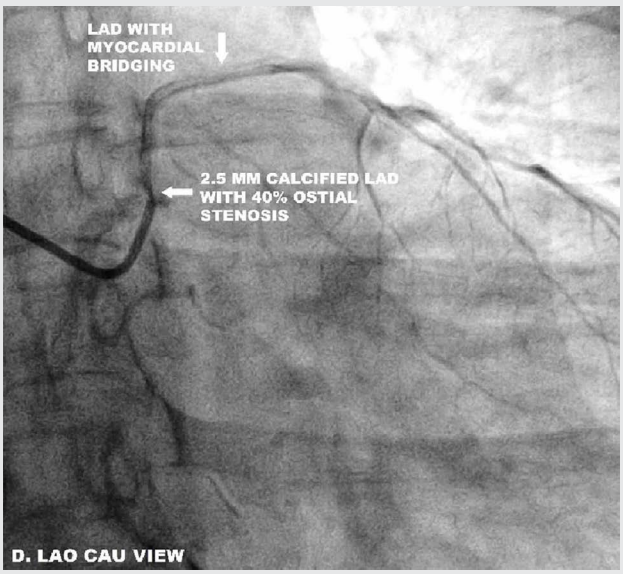
Figure 2: Coronary angiogram. A. LAO CAU view showing a very short single origin of the coronary arteries which immediately trifurcates to a 2.5 mm calcified LAD with 40% ostial stenosis with myocardial bridging; a 1.75 mm LCX with 70-80% ostial narrowing; and a 5 mm heavily calcified RCA with 99% ostial stenosis; B. LAO CRA view showing a very short single origin of the coronary arteries immediately trifurcating to LAD, LCX and RCA; C & D. LAO CAU view showing myocardial bridging; E. RAO CRA view showing 5 mm heavily calcified RCA.

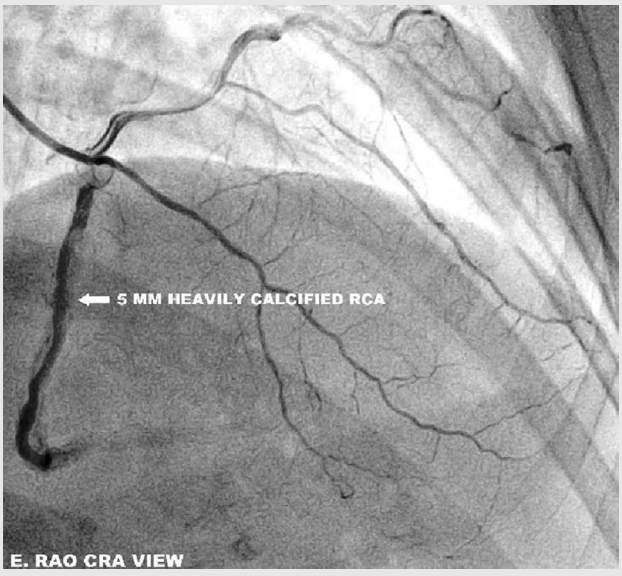
Legend: *LAO CAU, left anterior oblique caudal; LAO CRA, left anterior oblique cranial; RAO CAU, right anterior oblique caudal; LAD, left anterior descending artery; LCX, left circumflex artery; RCA, right coronary artery.*











DISCUSSION

Coronary anomalies often involve the origin or distribution of the artery. It affects a small percentage of the general population and are infrequently clinically significant.[5] It is commonly related to other cardiac abnormalities.[5] An SCA occurring without other major congenital anomalies has an incidence of 0.024–0.06%.[4, 6]

An SCA is a rare anatomic congenital abnormality with an incidence of <0.07% among patients undergoing coronary angiography; there is only one coronary artery originating from a single coronary ostium in the aortic sinus.[1-4, 7-8] Rather commonly occurring as an incidental finding, Angelini (2007) and Angelini et al., (1999) found that SCAs arising from the left aortic sinus are extremely more uncommon than one arising from the right, and finding all three coronary arteries originating from a single coronary ostium is very rare.[9-10]

Those with SCA are mostly asymptomatic at the time of diagnosis. Although some present with atypical chest pain and other nonspecific symptoms, which on further investigation reveal negative workup for ischemia, and/or no coronary artery disease (CAD).[10] Likewise, a few also present with typical chest pain, syncope, palpitations, ventricular tachycardia, myocardial infarction, and sudden cardiac death during physical exertion.[11] Our patient presented with hypotension that was refractory to fluid resuscitation which required vasopressor support. Additionally, she had no chest pain, palpitations nor dyspnea.

Although imaging modalities like echocardiography, coronary CT angiography and cardiac MRI are non-invasive strategies, invasive coronary angiography continues to be the gold standard for diagnosing and classifying SCAs.[12] Our patient had no significant findings on baseline cardiac workup before the elective BKA. Repeat TTE showed moderate global hypokinesis with a drop in LVEF, with significantly elevated cardiac biomarkers; invasive coronary angiography revealed an incidental SCA.

In 1979, Lipton proposed a way of classifying SCA angiographically (see Table 1) noting the origin of the ostium from the sinus of Valsalva (indicated by the letters R or L), anatomical distribution and course of the vessel (reflected by roman numerals I, II, or III), and the course of the transverse trunk (reflected by the and letters A, B, P, S, and C).[8] The RI and LI types usually follow a benign course, while R/LIIB or RIII types are more prone to cardiovascular morbidity and mortality, owing to the atypical course of the coronaries in between the outflow tracts.[13] And in 2005, Rigatelli et al. proposed a new classification system of the single coronary anomaly based on the clinical significance: class I – generally benign; class II – closely correlated with fixed myocardial ischemia, class III – significantly associated with sudden cardiac death; and class IV - linked with superimposed CAD.[14]

In a study by Turkmen et al. (2014), of the 215,140 patients undergoing invasive coronary angiography between 1998 to 2013, the prevalence of SCA was 0.031% with an incidence of 0.014 to 0.066%. Furthermore, by angiographic classification, prevalence was as follows: 9% type R-I, 34% type R-II, 15% type R-III, 24% type L-I and 18% type L-II.15 The three vessels traveled in between the aorta and pulmonary trunk in our patient, hence would be classified as the RIIIB type (see Figures 2A and 2B) by Lipton and class IV by Rigatelli.

Management strategies include plain medical, or complex revascularization: PCI with stenting or surgical CABG. Since most of these patients are asymptomatic, the majority can be managed conservatively by aggressive control of risk factors.[13] For those symptomatic patients, a multidisciplinary approach may be considered to determine the best management. For those with significant obstructive CAD, intervention through either of the two complex revascularization procedures is often necessary.[13]

Aside from the findings in the invasive coronary angiogram (1. SCA, and 2. significant ostial lesions in the LCX and the RCA), and a high Syntax score with lesser mortality favoring CABG as the procedure of choice (26.8% vs. 44%, p<0.001), surgical revascularization through CABG was advised, however the patient refused. The discussion was followed though a mutlidisciplinary team meeting, where the team acknowledged and respected the patient's wish. She was sent home comfortable after the vasopressors were discontinued with guideline-directed medical therapy and advice of close follow up. She likewise refused further renal replacement therapy at the outpatient which eventually led to her demise two weeks after discharge.

CONCLUSION

Cardiologists should be cognizant of the rare coronary anomalies with an increased risk of sudden cardiac death as it has implications for planning out definitive management. A multidisciplinary team approach in determining the best management strategy to employ is ideal. Guideline-directed medical therapy with aggressive control of risk factors are conservative measures that benefit patients with this type of incongruity.

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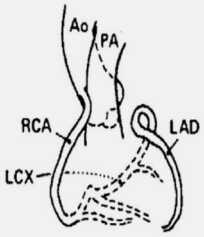
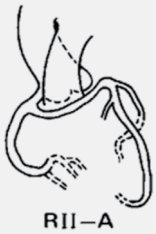
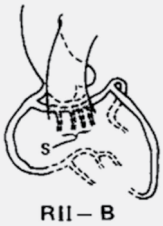
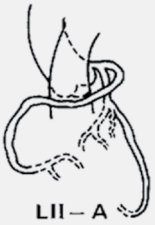
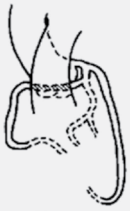
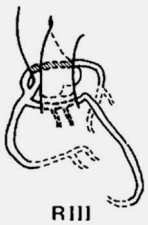
Table 1. Lipton's Classification of Angiographic Types of Isolated Single Coronary Artery Patterns.*[9, 14]

Originating from the right cusp and diagram	Description	Originating from the left cusp and diagram
 RI	Coming from the right coronary sinus, RI follows the course of the normal right coronary artery and normally gives off the posterior descending artery terminating as the posterolateral left ventricular branches. It differs from right dominant coronary system to reach the anterior basal surface of the heart as it pierces the atrioventricular groove giving off the anterior descending artery as its terminal branch. LI is a single coronary artery arising from the left sinus. It gives off the anterior descending branch terminating as the left circumflex branch. It differs from the usual left dominant system in that it continues beyond the crux into the right atrioventricular groove to supply marginal branches to the right ventricle.	 LI
 RII–A  RII–B  RII–P	The single coronary artery arises from the right or left coronary sinus, from this a very large trunk crosses the base of the heart to arrive in the vicinity of the normal contralateral coronary artery. Six subgroups can be distinguished, based on the site of origin and the route of this large transverse trunk. • RII-A: the single coronary artery originates from the right sinus, and the transverse trunk passes anterior to the conus of the right ventricle or the main pulmonary artery; • RII-B: the transverse trunk courses between the aorta and main pulmonary artery. It may be distinguished from the other types by identifying septal branches which arise from the transverse trunk as it crosses the superior aspect of the interventricular septum; or • RII-P: the transverse trunk courses posterior to the aortic root to arrive near the left coronary sinus at the atrioventricular groove. Similarly, single coronary arteries arising from the left coronary sinus may give off: • LII-A: a proximal branch which passes either anterior to the pulmonary trunk; • LII-B: between the aorta and pulmonary trunk; or • LII-P: posterior to the aorta. When the transverse trunk arrives near the right sinus of Valsalva, it courses in the atrioventricular groove in a manner similar to the normal right coronary artery.	 LII – A  LII – B  LII – P
 RIII	The single coronary artery originates from the right sinus of Valsalva, the circumflex branch and the anterior descending branch may arise separately from a common trunk, classified as RIII. The circumflex branch passes the atrioventricular groove retroaortically, while the anterior descending branch courses between the aorta and pulmonary trunk to the interventricular sulcus.	

Legend: *R - right, L - left, RCA - right coronary artery, LCA - left coronary artery, LAD - left anterior descending coronary artery, LCx - left circumflex coronary artery, Ao - aorta, PA - main pulmonary artery, A - anterior, B - between, and P - posterior to the great vessels. Shading of thetransverse trunkindicates this arterycourses posterior to the aorta.*

**Grouping, description and diagrams lifted from: 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; and Elbadawi A, Baig B, Elgendy I, et al. Single coronary artery anomaly: a case report and review of literature. Cardiol Ther. 2018 Jun; 7(1): 119-123. Doi: 10.1007/s40119-018-0103-4.*

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