Single coronary artery arising from the right sinus of Valsalva presenting with chest pain

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Radiology Case. 2016 Dec; 10(12):1-6 :: DOI: 10.3941/jrcr.v10i12.3016

ABSTRACT

Single coronary artery is a rare anomaly, with a reported incidence of 0.024%. It can be diagnosed on coronary computed tomography angiography, where the presence of ostial narrowing and compression between the great vessels must be carefully excluded, since these malignant forms can lead to sudden cardiac death and other ischemic complications. We describe a case of single coronary artery arising from the right sinus of Valsalva presenting with symptoms of ischemic chest pain.

CASE REPORT

CASE REPORT

A 48-year-old woman with hypertension presented with typical anginal chest pain for a week. No ECG changes were noted. Her serum lipid levels were mildly elevated, with a total serum cholesterol of 6.68 mmol/L (optimal level < 5.20 mmol/L), high density lipoprotein level of 0.90 mmol/L (optimal level > 1.00 mmol/L) and low density lipoprotein level of 4.94 mmol/L (optimal level < 2.60 mmol/L). Framingham risk score was low, calculated at 2.5.

CT coronary angiography (CTCA) was performed using a 320-slice single-source scanner (Aquilion ONE; Toshiba Medical Systems Corporation, Tokyo, Japan) with an injection of 60 ml of a low-osmolar contrast agent, iohexol (Omnipaque 350, GE Healthcare Inc.) at 4.5 ml/s. The scan was performed as a prospective, ECG-gated study.

This revealed a single coronary artery arising from a single ostium at the right coronary sinus, branching into a right coronary artery and left coronary artery shortly after the origin. The left coronary artery formed a long transverse trunk crossing the base of the heart and passing anterior to the conus of the right ventricle, (Figure 1) before branching into a small vessel with a marginal artery course and a normal caliber circumflex artery. The left anterior descending artery was absent. It was classified as a subgroup R II-A type anomaly [1-3]. A spot of calcified plaque was seen in the distal right coronary artery producing minimal stenosis (Figure 2), otherwise the entire right coronary artery was patent. No atherosclerotic plaque was seen in the left coronary artery, circumflex artery or its major branches. No ostial luminal narrowing was seen at the origin of the single coronary artery. The single coronary artery did not have a proximal intramural aortic course, which was carefully excluded on sagittal reconstructions. Hence, it was postulated that her anginal chest pain may have been caused by the absence of a left anterior descending with a very small caliber neighboring branch vessel in the course of the marginal artery, resulting in coronary insufficiency.

An elective dobutamine stress echocardiogram was performed, which was negative for ischemia. She declined conventional catheter angiography, so a decision was made to initiate medical management with antianginal and antihypertensive medication. The patient was subsequently lost to follow-up.

DISCUSSION

Etiology & Demographics:

An aberrant origin of the main stem coronary artery from the right sinus of Valsalva is known as one of the rarest coronary vessel anomalies and the incidence of single coronary artery is rarer still, reported at 0.024% of the population. Single coronary artery is a congenital anomaly defined as a solitary coronary artery arising from a single coronary ostium in the aortic trunk and supplying the whole heart [1, 4]. There is no known gender predisposition.

Clinical & Imaging findings:

The single coronary artery anomaly may be sub-grouped into three categories, according to Lipton's classification [1]. This classification, described by Lipton et al. in 1979, was based on conventional catheter angiographic findings and are summarized below:

- Group 1, where the solitary vessel follows the course of either a normal right or left coronary artery.
- Group 2, where the single coronary artery arises from the right or left coronary sinus and from this a large trunk crosses the base of the heart to arrive at the normal contralateral coronary artery. This is further subdivided into six subgroups depending on the site of origin and route of the large transverse trunk.
- Group 3 have a circumflex branch and anterior descending branch arising separately from a common trunk. The circumflex usually passes in a retroaortic route while the anterior descending branch courses between the aorta and pulmonary trunk to the intraventricular sulcus.

Further classification of these anomalies into malignant and benign forms depend on several factors, such as an interarterial course between the great vessels, proximal intramural course within the aorta, and ostial narrowing at the origin of the artery. Lipton's angiographic classification is summarized in schematic form below (Figure 3).

Treatment & Prognosis:

Single coronary arteries are usually asymptomatic, but some variants have been reported to carry a risk for serious cardiac events such as sudden cardiac death and myocardial infarction [5]. Even without an interarterial course, we postulate that the single coronary anomaly may be associated with ischemic symptoms if there are narrowed or absent, or if hypoplastic branches are in place of the usual normal caliber major vessels, such as the left anterior descending artery. Hypoplastic coronary arteries in isolation are known to be associated with sudden cardiac death and myocardial ischemia [6, 7]. It was first described in 1970 and is defined as a congenital underdevelopment of one or more major branches of the coronary arteries with significantly decreased length or diameter [8]. In a large study of coronary artery anomalies in patients undergoing catheter coronary arteriography, Lipton types R-I, R-II, R-III and L-I and L-II were identified as being potentially serious [9].

The treatment for single coronary artery has not been clearly established, but as general rule, surgery is considered for patients under 30 years of age with documented ischemia and without atherosclerotic disease [10]. Surgical management may include re-implantation of the origin of the anomalous coronary artery into a different coronary sinus, or coronary artery bypass grafting which can be performed to improve coronary flow distal to a compressed or narrowed anomalous vessel [10]. A conservative approach with medical therapy to reduce ischemia and avoidance of strenuous exercise has also been described [8, 10].

Differential diagnosis:

The main differential diagnosis for single coronary artery is absence or hypoplasia of one of the major coronary arteries, either the left anterior descending coronary artery and left circumflex coronary artery, or the right coronary artery. This should be carefully excluded during non-invasive coronary imaging as well as conventional catheter angiography if single coronary artery is suspected.

TEACHING POINT

Single coronary artery is a rare congenital anomaly which can have a benign or malignant clinical presentation, depending on the presence of luminal narrowing at the single coronary ostium or compression along its route. Even without an interarterial course, the anomaly may be associated with ischemic symptoms, particularly if there are absent or hypoplastic branches in place of the usual normal-caliber major vessels such as the left anterior descending artery.

REFERENCES

1. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated Single Coronary Artery: Diagnosis, Angiographic Classification, and Clinical Significance. Radiology. 1979; 130(1):39–47. PMID: 758666

2. Dodd JD, Ferencik M, Liberthson RR, Cury RC, Hoffmann U, Brady TJ, et al. Congenital Anomalies of Coronary Artery Origin in Adults: 64-MDCT Appearance. Am J Roentgenol. 2007; 188(2):W138–46. PMID: 17242219

3. Corbett M, Powers J, King S, Quinn M, Harris D Single Coronary Artery. J Am Coll Cardiol. 2009; 53(5):455. PMID: 19179205

4. Kariofillis, P., Mastorakou, I. & Voudris, V. Images in intervention. Origin of right and left coronary arteries from the right sinus of Valsalva as a common coronary trunk. JACC. Cardiovasc. Interv. 2009; 2, 805–6. PMID: 19695552

5. Frescura, C. et al. Anomalous origin of coronary arteries and risk of sudden death: A study based on an autopsy population of congenital heart disease. Hum. Pathol. 1998; 29, 689–695. PMID: 9670825

6. Amabile N, Fraisse A, Quilici J. Hypoplastic coronary artery disease: report of one case. Heart. 2005; 91(2): e12. PMID: 15657202

7. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. J Am Coll Cardiol. 1992; 20(3):640–7. PMID: 1512344

8. Evrengul, H., Ozcan, E. E., Turhan, H. & Ozturk, A. Single coronary artery originating from the right sinus of Valsalva and hypoplastic left anterior descending artery: An extremely rare combination of congenital coronary artery anomalies. Exp. Clin. Cardiol. 2012; 17, 243–4. PMID: 23592944

9. Yamanaka, O. & Hobbs, R. E. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet. Cardiovasc. Diagn. 1990; 21, 28–40. PMID: 2208265

10. Akcay, A. et al. Isolated Single Coronary Artery. Circ. J. 2008; 72, 1254–1258. PMID: 18654009

FIGURES

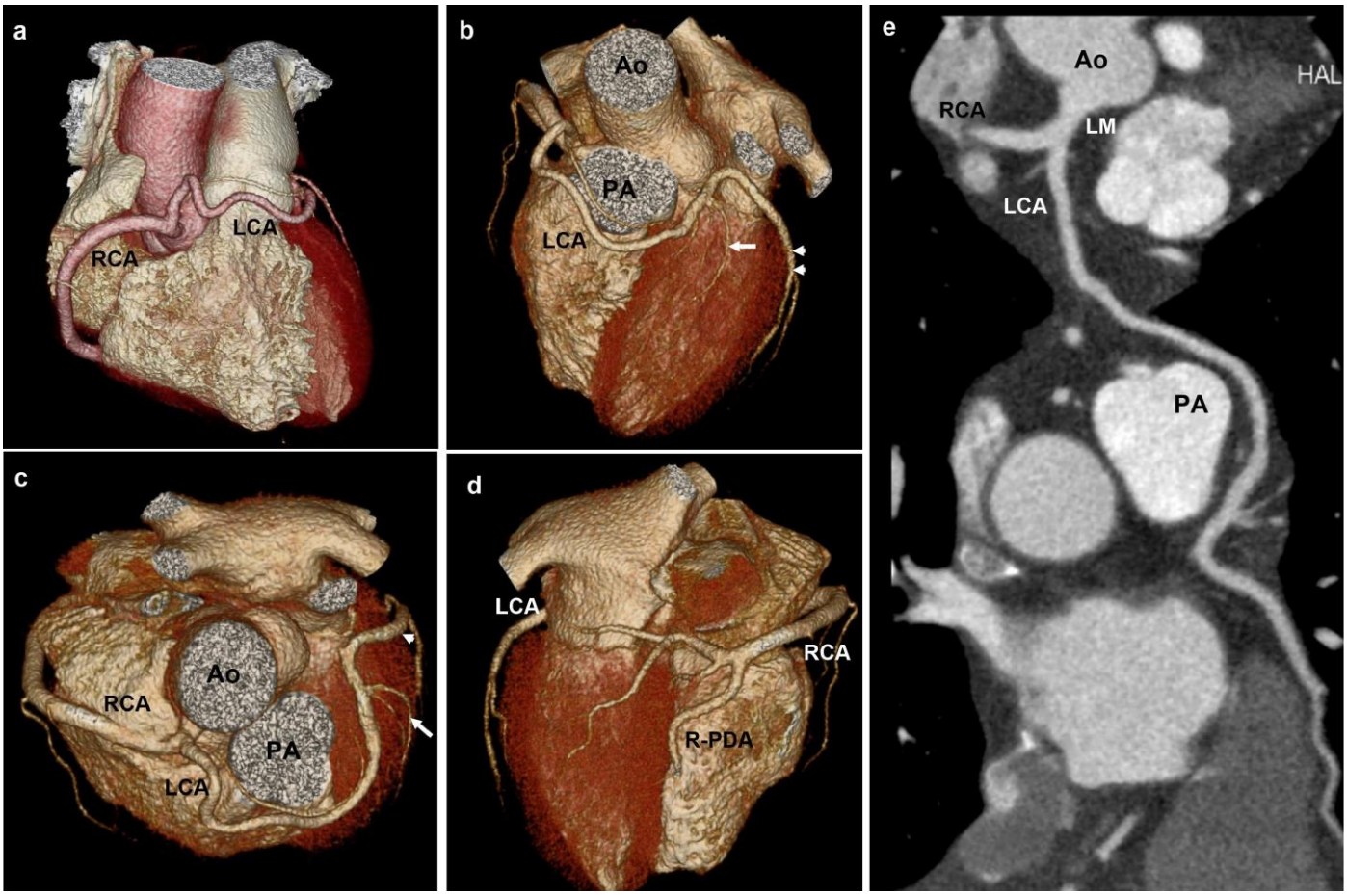

Figure 1: A 48-year-old woman with single coronary artery arising from the right sinus of Valsalva.

FINDINGS: Three-dimensional volume-rendered reformations Figure 1A: frontal view demonstrating the left coronary artery (LCA) and right coronary artery (RCA) arising from a single ostium from the right coronary cusp. Figure 1B: left anterior oblique cranial view and Figure 1C: left anterior cranial view shows the LCA continuing anterior to the right ventricular outflow tract before branching into a marginal artery (arrow) and circumflex artery (arrowheads). The left anterior descending artery was absent. Figure 1D: posterior view demonstrates the right coronary artery forming the right posterior descending artery (R-PDA). Figure 1E: curved planar reformations demonstrate the long transverse course of the left coronary artery anterior to the right ventricular outflow tract and pulmonary artery (PA). Aorta (Ao).

TECHNIQUE: contrast CT with three-dimensional volume-rendered reconstruction performed using a 320-slice single-source scanner (Aquilion ONE; Toshiba Medical Systems Corporation, Tokyo, Japan) with an injection of 60 ml of a low-osmolar contrast agent, iohexol (Omnipaque 350, GE Healthcare Inc.) at 4.5 ml/s. (ECG-gated, prospective gating, helical scan; A bolus-tracking method (monitoring level: carina; monitoring delay: 5 seconds; placement of ROI: descending aorta and HU threshold:180) was used to time the scan acquisition trigger;1.8 sec rotation time; pitch 0; 120 kV, 438 mA; slice thickness 0.5 mm)

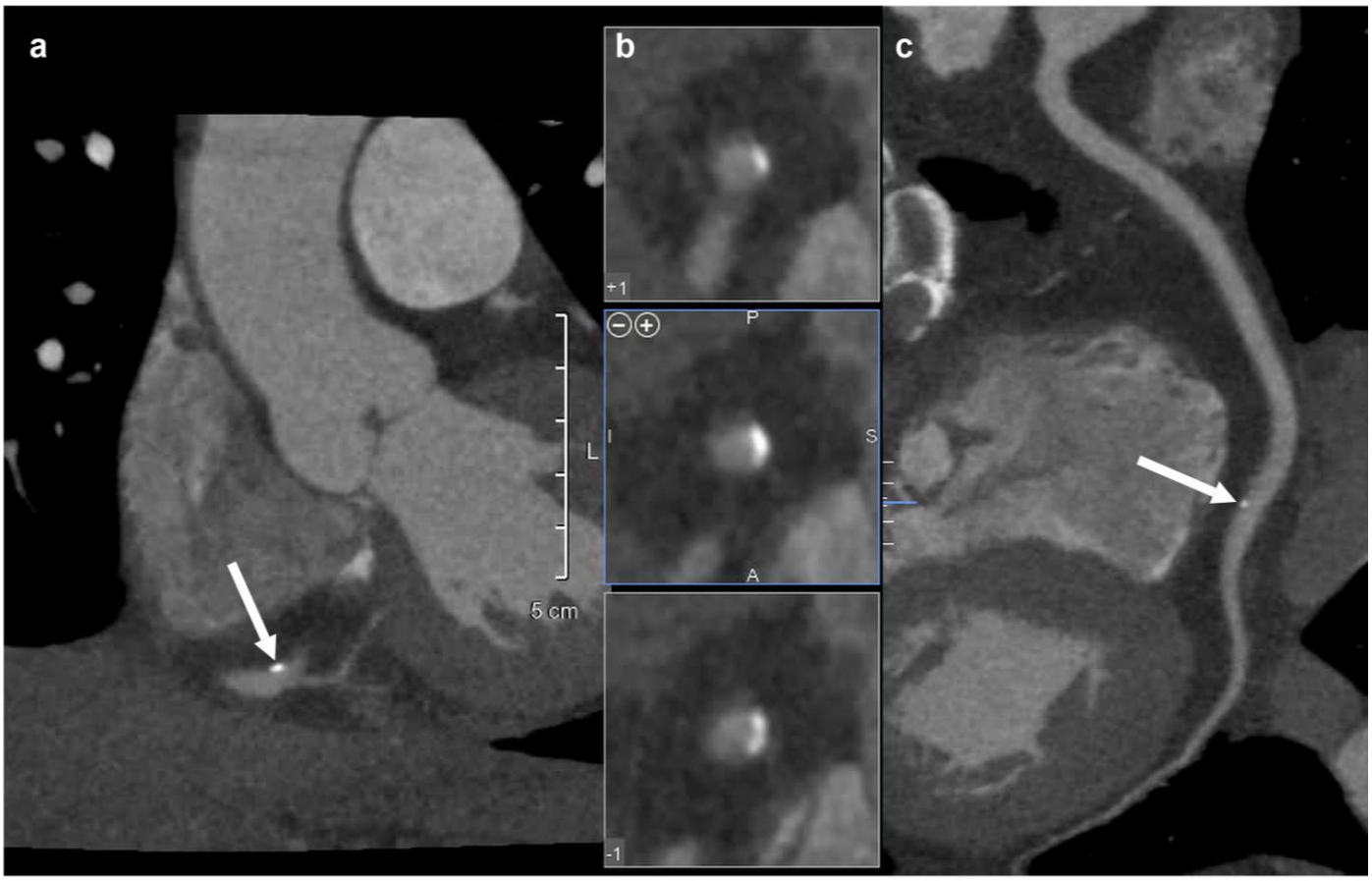

Figure 2: A 48-year-old woman with single coronary artery arising from the right sinus of Valsalva.

FINDINGS: Contrast CT with coronal multiplanar and curved-multiplanar reformations. Figure 1A: coronal multiplanar reformation demonstrating the distal right coronary artery containing a spot of calcified plaque (arrow). Figure 1B: cross section of the right coronary artery at the level of the calcified plaque revealing minimal stenosis. Figure 1C: curved-multiplanar reformation demonstrating the spot of calcified plaque (arrow) in the distal right coronary artery. The rest of the right coronary artery was patent.

TECHNIQUE: contrast CT with coronal and curved-multiplanar reconstruction performed using a 320-slice single-source scanner (Aquilion ONE; Toshiba Medical Systems Corporation, Tokyo, Japan) with an injection of 60 ml of a low-osmolar contrast agent, iohexol (Omnipaque 350, GE Healthcare Inc.) at 4.5 ml/s. (ECG-gated, prospective gating, helical scan; A bolus-tracking method (monitoring level: carina; monitoring delay: 5 seconds; placement of ROI: descending aorta and HU threshold:180) was used to time the scan acquisition trigger; 1.8 sec rotation time; pitch 0; 120 kV, 438 mA; slice thickness 0.5 mm).

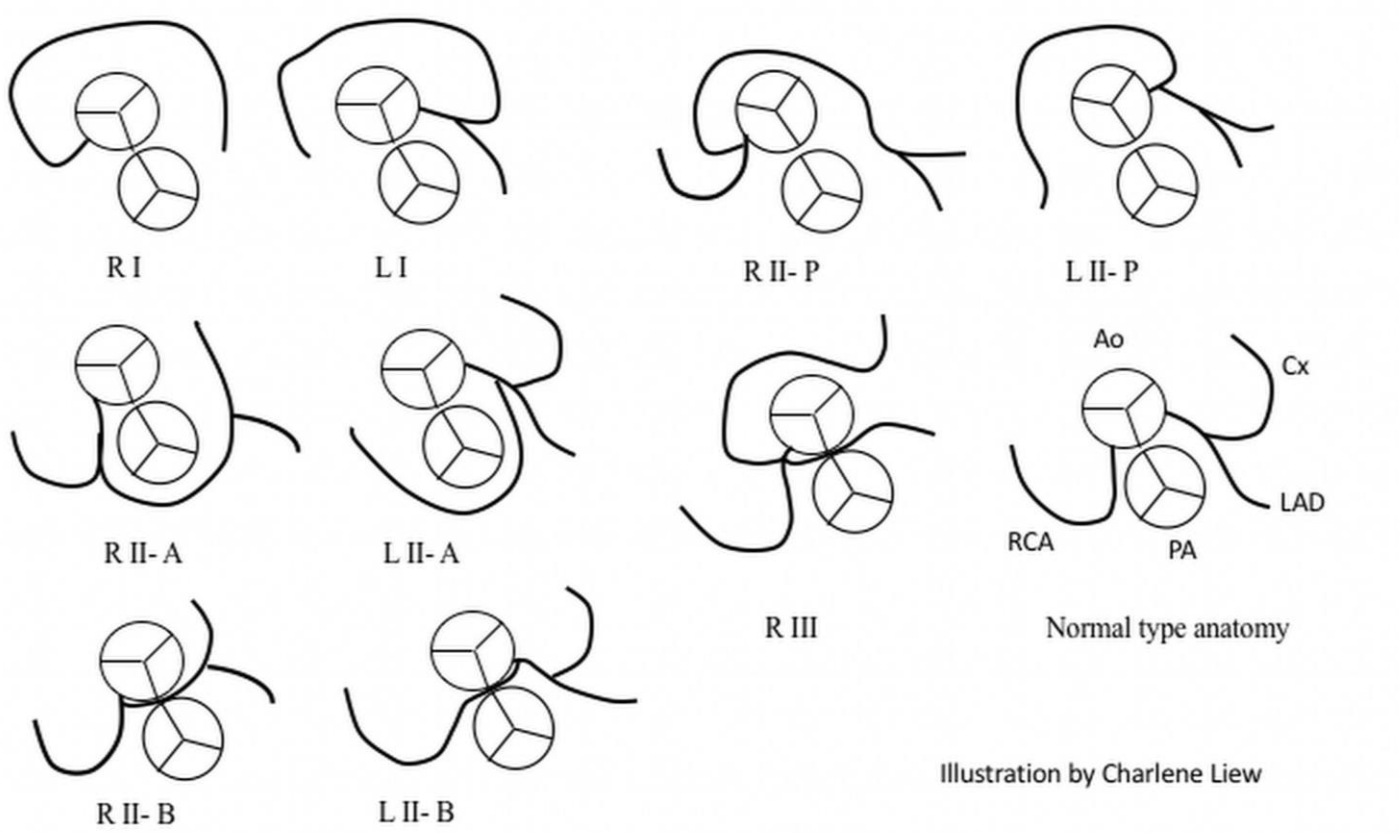

Figure 3: Schematic diagram representing Lipton's classification of single coronary artery.

The incidence of each anomalous subtype is as follows: R-I (0.0008%), R-II (0.015 %), R-III (0.004%), L-I (0.016%), L-II (0.009%) [9]. Ostial Location: R= Right Sinus of Valsalva, L= Left Sinus of Valsalva, Anatomical Distribution: I= Solitary dominant vessel follows the course of either a normal Right or Left coronary artery, II= One coronary artery arises from the proximal portion of the normally located other coronary artery, III= LAD and Cx arise separately from a common trunk originating from the right sinus of Valsalva, A= course of transverse trunk anterior to the great vessels, B= course of transverse trunk between the aorta and pulmonary artery, P= course of transverse trunk posterior to the great vessels, S= course of transverse trunk passes through the interventricular septum, C= combined type : a combination of diverse routes

	Classification	Description
Ostial Location	R	Right Sinus of Valsalva
	L	Left Sinus of Valsalva
Anatomical Distribution	I	Solitary dominant vessel follows the course of either a normal Right or Left coronary artery
	II	One coronary artery arises from the proximal portion of the normally located other coronary artery
	III	LAD and Cx arise separately from a common trunk originating from the right sinus of Valsalva
Course of the transverse trunk	A	Anterior to the great vessels
	B	Between the aorta and pulmonary artery
	P	Posterior to the great vessels
	S	Septal type passes through the interventricular septum
	C	Combined type : a combination of diverse routes

Table 1: Lipton's classification of single coronary artery.

Etiology	Congenital
Incidence	0.024% of the population
Gender ratio	No specific ratio
Age predilection	No specific age, usually asymptomatic until young adulthood or during exercise
Risk factors	No definite risk factor
Treatment	Surgical correction if presenting with ischemic symptoms, below the age of 30 and no concurrent atherosclerotic disease
Prognosis	Good, but certain subtypes carry risk for sudden cardiac death and myocardial infarction
Findings on diagnostic imaging	Computed tomography coronary angiography: A solitary coronary artery arising from a single coronary ostium in the aortic trunk and supplying the whole heart. Classification described in table 1.

Table 2: Summary table of single coronary artery

	CT findings
Single coronary artery	Solitary coronary artery arising from a single coronary ostium in the aortic trunk and supplying the whole heart.
Hypoplastic left coronary artery	Small caliber left main trunk, left anterior descending, left circumflex, normal caliber right coronary artery.
Hypoplastic right coronary artery	Small caliber right coronary artery, normal caliber left coronary artery.

Table 3: Differential diagnoses table for single coronary artery

ABBREVIATIONS

Ao = aorta
CTCA= computed tomography coronary angiography
Cx = circumflex artery
ECG = electrocardiogram
LCA = left coronary artery
PA = pulmonary artery
RCA = right coronary artery
R-PDA= right posterior descending artery

KEYWORDS

Cardiac imaging; Computed Tomography; Congenital anomaly; Chest pain; coronary vessels

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