# A Rare Single Coronary Artery Anomaly: Case Report and Review of the Relevant Literature

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**SUMMARY**

Single coronary artery is a rare congenital anomaly of the coronary arteries where only one coronary artery arises from the aortic trunk by a single coronary ostium. The clinical significance of single coronary artery is unknown. It is usually diagnosed incidentally during coronary artery angiograms or on postmortem evaluations. We report a case with the unusual feature of an isolated single coronary artery and present a review of the relevant literature.

**Key words:** *Coronary artery anomaly, single coronary artery (SCA), coronary angiography*

**ÖZET**

**Nadir Bir Tekli Koroner Arter Anomalisi: Olgu Sunumu ve Literatür Taraması**

Tekli koroner arterler, koroner arterlerin aort kökünde tek koroner ağızdan köken aldıkları nadir konjenital anomalilerdir ve klinik önemleri bilinmemektedir. Genellikle yapılan koroner anjiyografilerde ve otopsi örneklerinde rastlantısal olarak tespit edilmektedir. Bu yazımızda sağ koroner arter kaynaklı nadir bir tekli koroner arter anomali olgusundan ve konu ile ilgili literatür taramasından bahsettik.

**Anahtar kelimeler:** *Koroner arter anomalisi, tekli koroner arter, koroner anjiyografi*

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## Introduction

Single coronary artery (SCA) is a rare congenital anomaly originating in the aortic root through a single coronary ostium, supplying the entire heart (1). These anomalies are commonly an incidental finding during coronary angiography (CAG). The incidence of isolated SCA is only 0.0024–0.066% in the general population undergoing CAG (2). It is reported that 40% of SCA anomalies are associated with congenital heart diseases such as tetralogy of Fallot, transposition of great arteries, persistent truncus arteriosus, pulmonary atresia, coronary arteriovenous fistula and bicuspid aortic valve (3).

The prognostic significance varies, generally considered benign, ranging from exertional angina, myocardial ischemia to sudden death especially in young adults. Patients with the left coronary artery originating from the right coronary sinus have a high mortality rate before 20 years of age (~ 60%). Deaths occur usually during or after strenuous physical activity because of having a coronary artery branch following a dangerous course between the great vessels, as there is an increase in blood flow inside (4).

Nearly 15% of the patients may have myocardial ischemia directly caused by the abnormal anatomy of the arteries (4-7);

- Presence of an acute aortocoronary angulation, and a sinuous proximal course determining a pattern of turbulent flow and accelerated atherosclerosis,
- Coronary spasm resulting from its torsion movement,
- A narrowed slit-like orifice that collapses in a valve-like manner, thereby limiting the blood flow,
- Within the aortic wall, proximal intramural course of the anomalous vessel, which is squeezed,
- The compression of the anomalous vessel along its course between the aorta and the pulmonary artery.

In patients with SCA, the incidence of sudden death is low and the mechanisms are not fully understood. Long term myocardial ischemia, which may result in sparse fibrosis in the myocardium and predispose to lethal ventricular arrythmias, is the most accused phenomenon (8).

Lipton, *et al* classified SCAs into nine patterns according to the site of origin and anatomical distribution of the branch of the coronary artery (Table I-II).

## Case report

A fifty-six-year old female with hypertension, type 2 diabetes and a history of smoking has applied to our cardiology department with dyspnea and substernal chest pain intensifying with exercise. Physical examination was normal. Electrocardiography showed nonspecific ST-T changes in leads V1-V6. Echocardiography showed normal left ventricular morphology and no abnormal regional wall motion with ejection fraction of 60%. Exercise treadmill test (Bruce's protocol) was performed that showed significant ST-depression in precordial leads (V4–V6) in stage 2. Coronary angiography was planned for further evaluation and performed via the right femoral approach. Several attempts for left coronary cannulation were failed. Right coronary injection showed a SCA arising from the right sinus of Valsalva (Figure 1A). The absence of a left coronary artery confirmed by aortography. Our case was classified as "R-III-C" pattern - a rare anomaly of coronary arteries (Figure 1B). The SCA originated from the right sinus of Valsalva, and giving off branches for the LAD and LCX seperately from the proximal part (Figure 2). Course of the transverse trunks was combined type-LAD is anterior and LCX is posterior to great vessels.

## Discussion

Single coronary artery is a rare anatomical condition that may be encountered during coronary angiography. Unfortunately, there is still not a consensus on the management of SCA. Long-term data about medical management and the surgical approach are lacking. Cardiac surgeons and interventional cardiologists should be aware of these pathologies because recognition of them is mandatory for appropriate management and prevention from inadvertent transecting or ligaturing during surgery.

Once identified, participation in sports should be prohibited. Surgical correction, when feasible particularly in young patients, can restore coronary blood flow (9). It is suggested that medical treatment is usually adequate for SCA in middle age and elderly patients in the absence of ischemia and/or acute coronary syndrome. The surgical indication for asymptomatic elderly patients is not clear because, at older ages, the arteries are less compressible and unless there is concomitant obstructive coronary disease the surgery is not routinely recommended (10).

The surgical approach may involve decompression of its course or reimplantation of the anomalous vessel in the correct coronary sinus. In addition, coronary artery bypass grafting alone can be used as the standard procedure for restoring normal distal coronary flow, especially whenever an interarterial coronary artery is detected. Previous studies suggest

| Table I. Classification of angiographic types of single coronary artery (Modified Lipton's Classification) | |
|---|---|
| *Code* | *Description* |
| Ostial location | |
| R | Right Valsalva Sinus |
| L | Left Valsalva Sinus |
| Anatomical distribution | |
| I | Solitary dominant vessel follows the course of RCA or LCA (R-I or L-I) |
| II | One coronary artery arises from the proximal part of the normally located other coronary (R-II or L-II) |
| III | LAD and LCX arise separately from common trunk originating from the right Valsalva sinus (R-III) |
| Course of the transverse trunk | |
| A | Anterior to great vessels (Anterior to right ventricul) |
| B | Betwenn the aorta and pulmonary arteries |
| P | Posterior to the great vessels |
| S | Septal type: part of the route passes through interventricular septum |
| C | Combined type: combination of diverse routes |

*RCA*, right coronary artery; *LCA*, left coronary artery; *LAD*, left arterior descending coronary artery; *LCX*, left circumflex coronary artery.
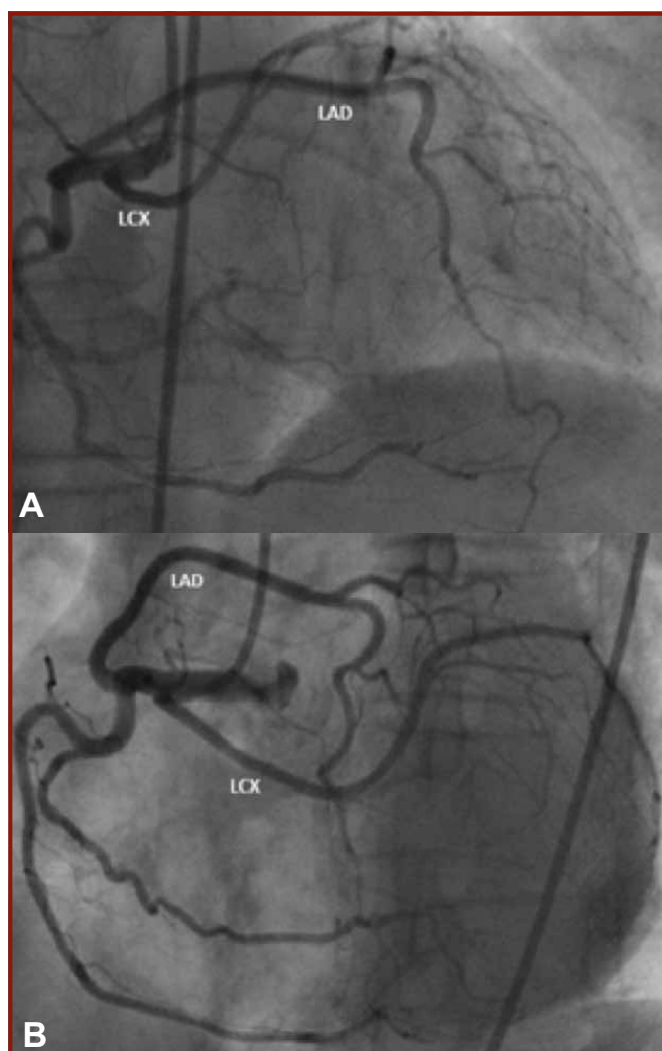
**Figure 1.** Coronary angiography in right-caudal position demonstrates a single coronary artery arising from right sinus of Valsalva **(A)**. Anomalous origin of the LAD and LCX from right sinus of Valsalva **(B)**. **LCX**: Left Circumflex Coronary Artery; **LAD**: Left Anterior Descending Artery.
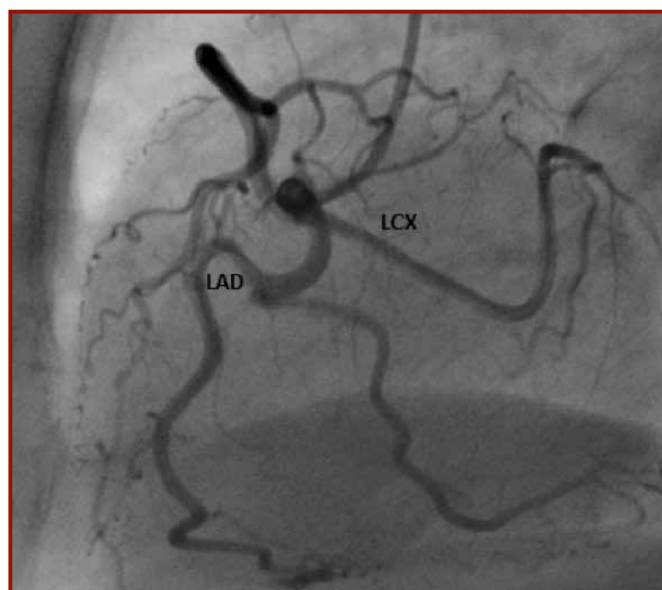


**Figure 2.** Coronary angiography in right-cranial position demonstrates a single coronary artery arising from right sinus of Valsalva.

that the surgical ligation of the anomalous coronary artery may be beneficial because it avoids competitive flow from the graft and native coronary artery (11). The surgical treatment was preferred because coronary angioplasty could compromise the other important branches. Nevertheless, new advancements in coronary interventional devices and techniques have made angioplasty another feasible therapeutic choice.

## Conclusion

Without crucial clinical data, it is undoubtedly difficult to set guidelines/recommendations for the management of SCA. The recognition and identification of this anomaly are of clinical importance because the symptoms cannot be differentiated from atherosclerotic coronary artery disease. Among low-risk female patients with chest pain and a positive stress test, coronary artery anomaly should be considered and an angiographic study should be performed.

Coronary anomalies are usually detected during coronary angiography, the gold standard method for the detection of SCAs, enabling simultaneous percutaneous interventions for treatment of associated or non-associated coronary lesions. However, X-ray angiography is limited by its inability to provide information regarding the spatial orientation of the anomalous coronary artery with regarding to the surrounding cardiovascular structures (12).

In addition to coronary angiography, transesophageal echocardiography and contrast-enhanced electron-beam tomography have also been recommended to detect the anomalous coronary artery, other structures and coexisting anomalies with their spatial orientation.

In our case, there were no atherosclerotic lesions in her coronary arteries. No concurrent congenital heart disease was diagnosed with transthoracic echocardiography, catheterization of the left heart, and ventriculography. The patient was managed with risk factor modification-advised to quit smoking and avoid strenuous physical activity in the absence of associated severe coronary artery disease and/or overt ishemia. She has been followed since 2009 without any symptoms.

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