Case report

# A very rare coronary artery anomaly: Twin circumflex arteries associated with acute coronary syndrome - two cases report

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**Abstract**

Circumflex coronary artery anomalies are the most common type so far observed. However, a dual origin of the circumflex is an extremely rare anomaly. We describe two different patients admitted to our clinic with acute coronary syndrome at the same day. Angiography revealed twin circumflex arteries: one from the left main artery and the other from the proximal right coronary artery.

**Keywords:** *twin circumflex coronary artery; acute coronary syndrome; coronary artery anomaly.*

## Introduction

The increasingly extended use of diagnostic coronary angiography is discovering numerous congenital anomalies of the coronary arteries. At first, they were considered simple angiographic findings and there was a tendency to characterize them as benign [1, 2]. However, this attitude was undermined by reports of cases of sudden death, acute myocardial infarction, angina and syncope associated with their presence [3, 4]. A dual origin of the circumflex artery (Cx) is an extremely rare anomaly. The incidence of such as coronary anomalies is relevant not only for educational attentions but, more importantly, for public health issues. We describe two different patients admitted to our clinic with acute coronary syndrome at the same day.

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## Case report

First case, a 65-year-old man with an eight hours history of chest pain was referred to our clinic. He had no history of coronary artery disease, and alcohol or drug use. The only risk factor for atherosclerosis was hypertension.

His physical examination, echocardiogram except left ventricular hypertrophy were all normal. The electrocardiogram showed ST-segment depression in leads V3-V6 and D1-aVL (Figure1A). Findings of blood tests were unremarkable except for increases in cardiac enzymes. The patient was transferred to our catheterization laboratory for diagnostic coronary arteriography.

Conventional angiography revealed no significant stenosis of the left anterior descending coronary artery (LAD) and the left Cx artery, however intermediate artery (IM) had critical stenosis (Figure 1B). Surprisingly, right coronary angiogram showed an additional Cx artery and a right coronary artery (RCA) arising separately from the right sinus of Valsalva (Figure 1C). Both of them had non obstructive plaques. Thereafter, stenting was successfully performed for the IM artery stenosis.

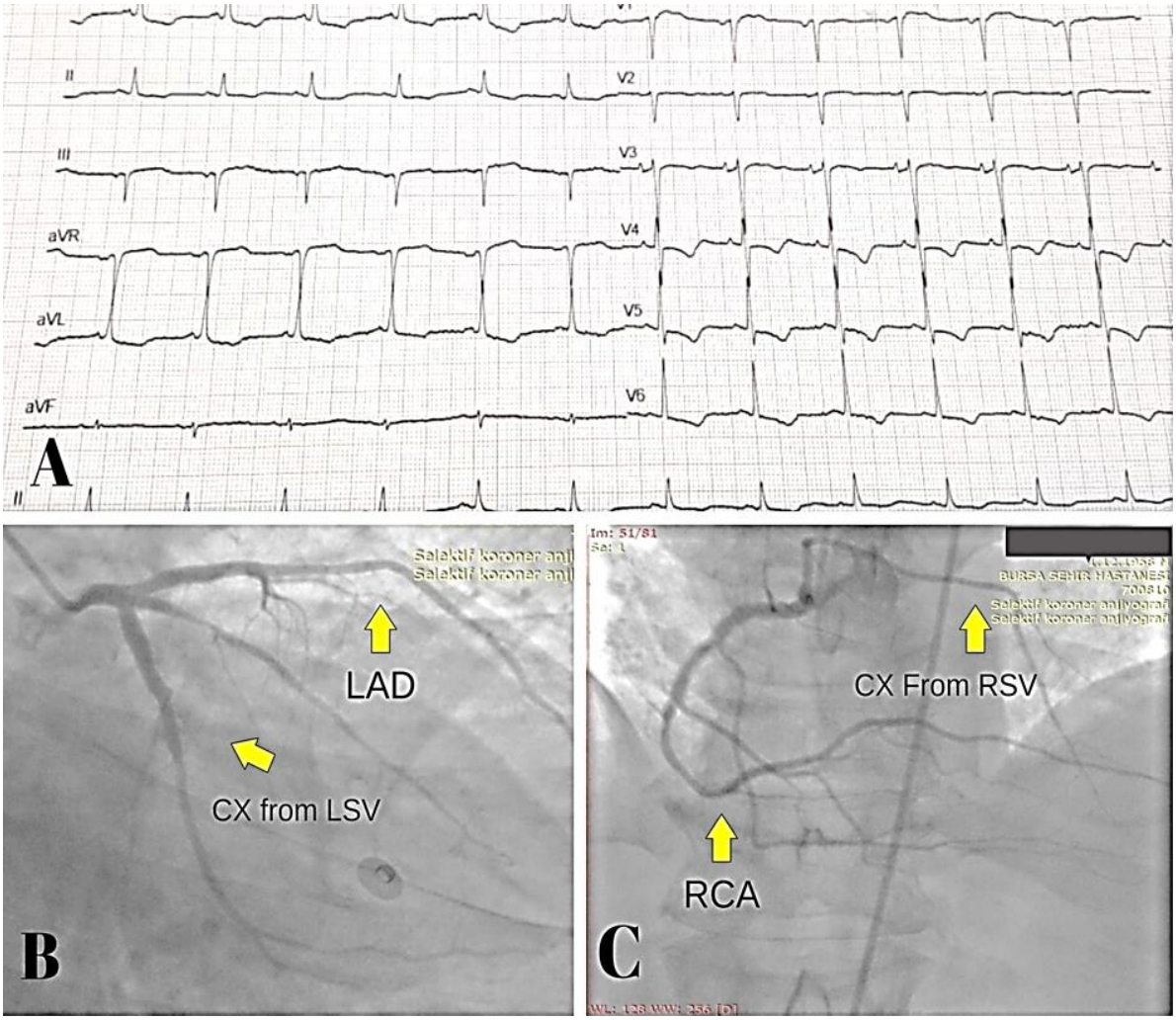


**Fig. 1. A.** The electrocardiogram showed ST-segment depression in leads V3-V6 and D1-aVL. **B**. Coronary angiogram in the right caudal view shows the left coronary artery, circumflex artery and intermediate artery. **C**. Coronary angiogram in the left anterior oblique cranial view shows the circumflex artery and right coronary artery originating from right sinus of Valsalva. *(CX: circumflex artery, IM: Intermediate artery, LAD: left anterior descending artery, LSV: Left sinus of Valsalva, RCA: Right coronary artery, RSV: Right sinus of Valsalva)*

Second case, a 49-year-old man had history of chest pain for four hours and high troponin levels was referred to our clinic. The only risk factor for atherosclerosis was smoking.

His physical examination, echocardiogram, and electrocardiogram (Figure 2) reports were all normal. Following physical examination and initial tests, a diagnostic coronary arteriography was performed.

Angiography revealed that no significant stenosis of the LAD and the left Cx artery (Figure 3A). Right coronary angiogram showed an additional Cx artery and a RCA arising separately from the right sinus of Valsalva (Figure 3B-3C). The right Cx artery had significant stenosis and performed stenting (Figure 3D).

The post-interventional period was in a good condition for both of the patients, and the patients was followed up for 3 days and discharged without any complications.

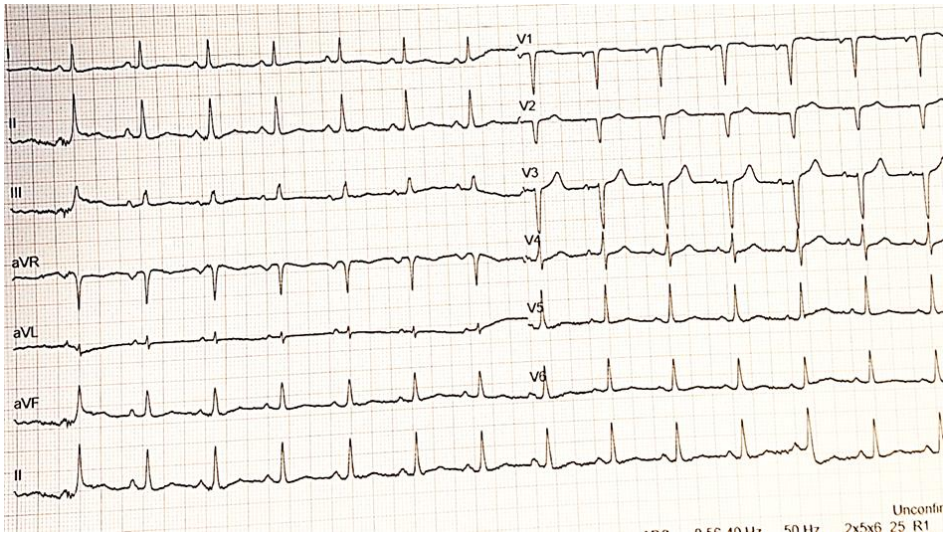


**Fig. 2.** The electrocardiogram showed no ST-segment depression or elevation

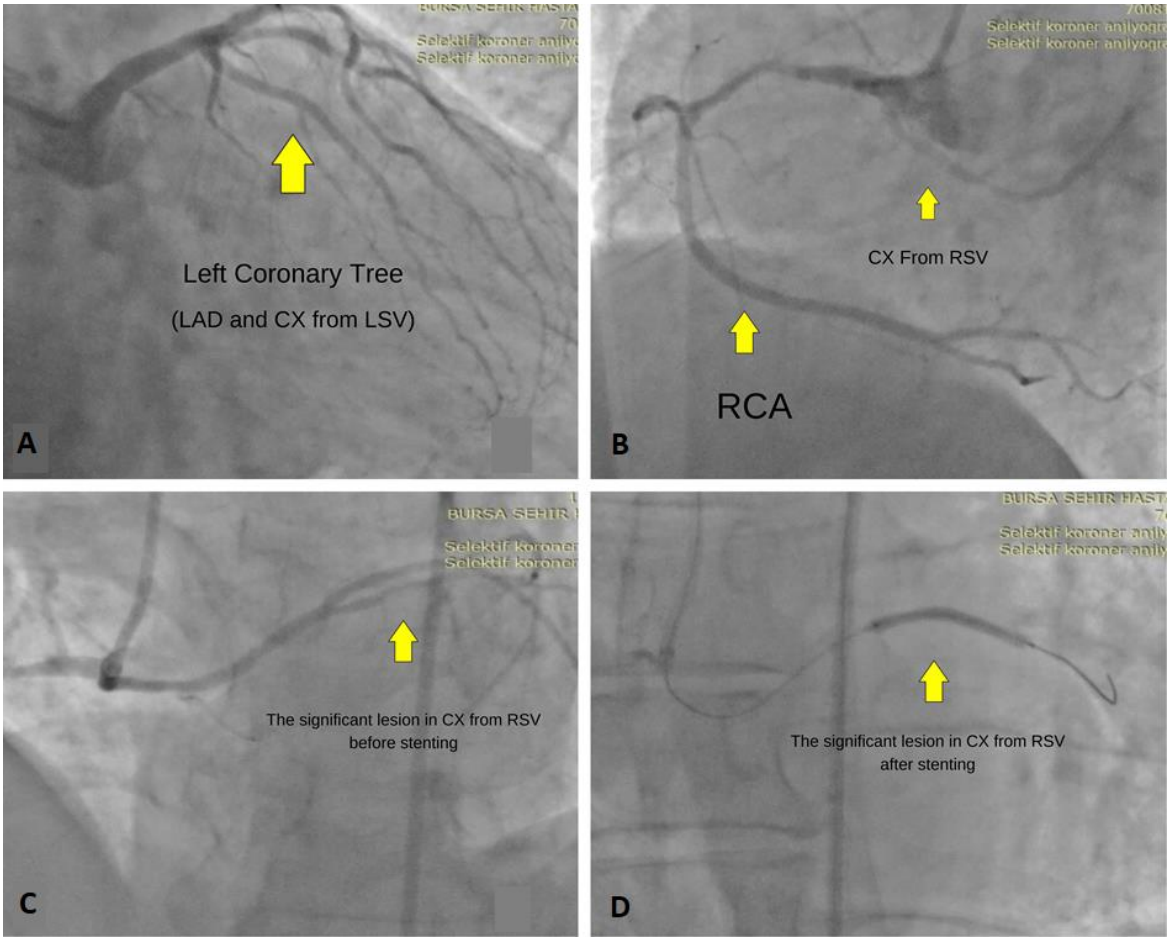


**Fig. 3. A.** Coronary angiogram in the left caudal view shows the left coronary artery tree with a circumflex artery. **B.** Coronary angiogram in the left anterior oblique cranial view shows the circumflex artery and right coronary artery originating from right sinus of Valsalva. **C.** The significant lesion, in the left anterior oblique cranial view, shows the circumflex originating from right sinus of Valsalva. D. After stenting the significant lesion, in left anterior oblique view shows the circumflex artery originating from right sinus of Valsalva *(CX: circumflex artery, LAD: left anterior descending artery, LSV: Left sinus of Valsalva, RCA: Right coronary artery, RSV: Right sinus of Valsalva)*

## Discussion

Coronary artery anomalies are found in 0.6-1.5% of coronary angiograms [1]. Although most of them have no clinical significance, they may cause acute myocardial damage and/or chronic injuries in the area supplied by the anomalous coronary artery arising from the incorrect coronary sinus of Valsalva [5]. Some coronary artery anomalies may cause chest pain, arrhythmia, heart failure and sudden death [6]. Myocardial ischemia can occur because of earlier and more aggressive atherosclerosis compared to a normal coronary artery [7]. That was found exclusively in anomalous vessels arising from the right side with a retroaortic course [8].

The most frequently found anomalies include a Cx artery with a separate ostium from the LAD originating in the left coronary cusp, an origin of the Cx artery taking off from the RCA or arising separately from the right coronary cusp [1]. This anomaly is thought to be benign and is usually clinically silent. However, it was observed in the Coronary Artery Surgery Study that the incidence of stenosis was greater in the Cx arteries originating from the right coronary sinus compared to normal Cx arteries originating from the left main coronary artery. There have been many reports of anomalous coronary arteries and their association with accelerated atherosclerosis resulting in myocardial infarction and sudden death, depending upon their origin, course, and termination [2, 9, 10]. The incidence of Cx anomalies is summarized in Table 1 [1, 8, 11].

**Table 1**. The incidence of CX anomalies

| Circumflex Artery Anomaly | Incidence (%) |
|---|---|
| Separate origin of LAD and CX in LSV | 0,7[1] |
| CX from RSV or RCA | 0,37[1] |
| Absent CX | 0,003[8] |
| Intercoronary communication (CX-RCA) | 0,002[11] |
| Congenital ostial stenosis or atresia of CX | 0,001[11] |
| CX - PA/RV fistula | 0,02[11] |
| Others | 0,2[1] |

*CX: Circumflex, LAD: Left anterior descending, LSV: Left sinus Valsalva, PA: Pulmonary artery, RCA: Right coronary artery, RV: Right ventricle*

Dual connection of the LAD to the left main coronary artery and the RCA is an extremely rare congenital coronary anomaly. This anomaly was classified into four types by Spindola-Franko et al. Double LAD type 1 is the most common. They found an incidence of about 1% for the type 1 in normal heart. Double RCA is also a very rare congenital coronary anomaly. There is not enough studies regarding double RCA in literature. Double RCA has been reported as case reports in only a few papers [12, 13].

The anomalous origin of the Cx artery from the proximal right coronary artery or from the right sinus of Valsalva was first described by Antopol and Kugalin 1933 [14]. There have been only 7 previously reported cases of bilaterally arising twin Cx arteries: one Cx artery arose from the left main coronary artery and the others from the right aortic sinus (3 cases) or the ostium of the right coronary artery (4 cases) following thereafter a retroaortic course to the left. In 2008 [15-19), Attar et al. reported a case of twin Cx arteries: one from the left main artery and the other one originated from the right coronary sinus [15]. Along the same lines, Van der Velden et al. presented a case with the coexistence of coronary fistulae and twin Cx arteries [16]. Elsewhere, Karabay et al. presented a case of twin Cx arteries arising from the left and right coronary systems with acute inferior myocardial infarction treated via percutaneous coronary intervention [5]. In a study by Cicek et al. there were significant stenoses at both of the twin Cx arteries, leading to heart failure [17]. Andreou et al. reported a case for preoperative identification of this anomaly in patients undergoing aortic valve

surgery [18]. Another investigation by Otlu et al. reported transradial percutaneous coronary intervention in a patient with twin Cx arteries [19]. In this case report, we described two patients with twin Cx arteries: one originating from the left main coronary artery and the other one, a Cx, arising from the proximal part of the right coronary artery. Additionally, one of our case had critical right Cx stenosis.

## Conclusion

The most important problem in diagnosing double Cx arteries the separate origin of the two Cx arteries from different ostia on the left or right aortic sinus of Valsalva. In the absence of significant stenosis in the normal Cx, an anomalous Cx arising from the RCA, right sinus of Valsalva, or aorta should be suspected in a patient with acute inferior, posterior myocardial infarction or acute coronary syndrome [5]. Thus, the angiographer must always keep in mind this possibility.

### Consent

Written informed consent was obtained from the patients for publication of these case reports. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

### Competing interests

The author(s) declare that they have no competing interests.

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