

# Double Right Coronary Artery Anomaly Coexistent with Double Coronary Artery Fistula: Case Report

## Çift Sağ Koroner Arter Anomalisi ile Birlikte Çift Koroner Arter Fistülleri

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**ABSTRACT** Coronary artery anomalies are rare cases and may be encountered incidentally during coronary angiography. Because of the relationship of these anomalies with hemodynamic disturbance, chest pain and sudden cardiac death, it is important to keep in mind the probability of coronary artery anomalies, particularly in young adults. Double right coronary artery (RCA) anomalies are usually benign conditions, while coronary artery fistulae may lead to volume overload or myocardial ischemia. To our knowledge, the case we present here is the first case of double RCA anomaly which coexists with double coronary artery fistula, in the available literature.

**Key Words:** Heart defects, congenital; coronary vessel anomalies; vascular fistula; heart failure; coronary angiography

**ÖZET** Koroner arter anomalileri nadir durumlar olup koroner anjiyografi sırasında tesadüfen saptanabilir. Bu anomalilerin hemodinamik bozukluk, göğüs ağrısı ve ani kardiyak ölüm ile ilişkisi nedeni ile özellikle genç erişkinlerde koroner arter anomalisi olasılığının akılda tutulması önemlidir. Koroner fistüller volüm yüklenmesi veya miyokardiyal iskemiye yol açabilirken çift sağ koroner arter (RCA) anomalileri ise genellikle benign durumlardır. Mevcut bilgimiz itibarı ile burada sunmuş olduğumuz vaka literatürdeki çift koroner arter fistülü ile birlikte çift RCA anomalisi olan ilk olgudur.

**Anahtar Kelimeler:** Kalp kusurları, doğumsal; koroner damar anomalileri; damar fistülü; kalp yetersizliği; koroner anjiyografi

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Coronary artery anomalies are uncommon entities which are present at birth but rarely symptomatic during childhood. Most of the cases have been determined incidentally during coronary angiography or autopsy. Some of these anomalies may be associated with hemodynamic disturbance, chest pain or sudden cardiac death particularly in young adults. Furthermore, to be aware of the presence of coronary artery anomalies prior to coronary angioplasty or cardiac surgery is crucial. Coronary artery fistula keep their importance as the a most common hemodynamically significant congenital coronary anomalies although they have a relatively low incidence.<sup>1</sup> However, double right coronary artery (RCA)

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anomaly is an extremely rare condition in comparison with coronary artery fistulae, and it doesn't cause any hemodynamic changes, unless accompanied by another anomaly. To the extent of our knowledge, so far, only twenty-eight cases of double RCA anomaly have been reported and present case is the first one case of double coronary artery fistula coexisting with double RCA anomaly.

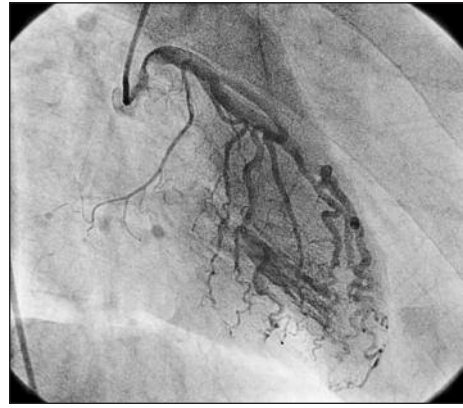
## CASE REPORT

A 44-years-old male patient was admitted to the cardiology outpatient clinic with a complaint of post-exertional squeezing chest pain lasting for the last three months. He had only smoking as a risk factor of coronary artery disease. Physical examination, heart rate, blood pressure, routine laboratory tests and ECG were within in normal ranges.

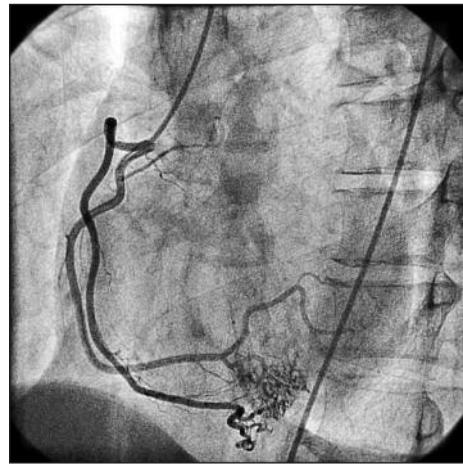
However, during the recovery phase of treadmill exercise stress test, ST segment depressions were seen in leads II, III and aVF. Coronary angiography revealed a fistula from distal left anterior descending artery (LAD) into the left ventricle (Figure 1). In addition, a double RCA (anterior and posterior) with single ostium originating from the right coronary sinus of Valsalva was also identified (Figure 2). Both RCAs were terminating as posterior descending arteries (PDAs), and there was another fistula from anterior RCA to the right ventricle (Figure 3). Doppler evaluation of the pulmonary to systemic flow ratio ( $Q_p/Q_s$ ) by transthoracic echocardiography was detected to be normal.

## DISCUSSION

According to the literature known until 1999, the incidence of coronary anomalies ranged between 0,3-1,3% for the people in general population undergone a cineangiogram for suspected obstructive disease.<sup>2-12</sup> However, Angelini and co-workers have reported a higher incidence (5,6%) than previous studies, and this discrepancy was explained as a result of their strict methodology, which was proposed and adopted by themselves.<sup>13</sup> The role of



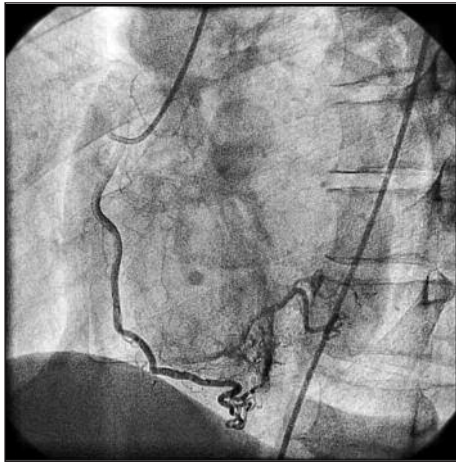
**FIGURE 1:** Angiography of left coronary system on right anterior oblique view demonstrating a fistula that involves the left anterior descending artery with drainage into the left ventricular chamber.



**FIGURE 2:** Angiography of right coronary system on left anterior oblique view demonstrating a double RCA with single ostium originating from right coronary sinus.

the coronary artery anomalies on premature cardiac morbidity and mortality may not be ignored despite its infrequent occurrence. In a study by Eckart et al. consisting of 126 nontraumatic sudden deaths in young adults, the most common cause of sudden death was an identifiable abnormality, especially coronary artery abnormalities ( $n=39, 61\%$ ).<sup>14</sup>

According to the modified version of classification system proposed by Angelini and co workers, coronary artery anomalies have been grouped into four main topics and these are anomalies of



**FIGURE 3:** Angiography of right coronary system on left anterior oblique view demonstrating a fistula that involves the posterior descending artery with drainage into the right ventricular chamber.

origination and course, anomalies of termination, anomalies of intrinsic coronary arterial anatomy and anomalous collateral vessels.<sup>15</sup> These anomalies may also be classified as hemodynamically either significant or insignificant.

Coronary artery fistula is defined as an abnormal communication between a coronary artery and a cardiac chamber, coronary sinus, superior vena cava or pulmonary artery. Therefore, coronary artery fistulae have been classified as anomalies of termination. In a recent study, the incidence of this anomaly is reported as 0,87% of all patients who undergo coronary angiography.<sup>13</sup> Right coronary system is the most frequent site of origin according to earlier studies, however more recent studies suggest that the left system may be the more common site.<sup>16,17</sup> Involved artery is usually dilated and tortuous. These anomalies may lead to significant hemodynamic disturbances as related to drainage site and volume.<sup>1</sup> Drainage site of fistula has the most important role in determining the hemodynamic disturbance. Most common drainage sites are right ventricle and pulmonary artery.<sup>16,17</sup> Drainage into the right-sided cardiac chambers may lead to a left-to-right shunt, whereas drainage into the left-sided cardiac chambers hemodynamically resembles the aortic regurgitation. Clinical symptoms are related to the hemodynamic abnormality.<sup>18</sup> This condition

may lead to myocardial ischemia as a result of steal phenomenon at the site of myocardium that is supplied by fistulous artery. Cardiac catheterization is still the gold standard for the diagnosis of coronary artery fistulae. It is able to demonstrate the anatomy, size, number, origination and termination site of fistulae. Magnetic resonance imaging (MRI) is a good alternative for imaging of proximal coronary abnormalities but it may be less effective on distal course imaging. Recently, multidetector row computed tomography (MDCT) cardiac imaging has given great results for distal coronary artery and side branch imaging.<sup>19</sup>

Double RCA anomaly is extremely rare among the congenital anomalies of the coronary arteries. The first double RCA anomaly was described by Barthe et al. in 1994.<sup>20</sup> To our knowledge, a total of 28 double RCA anomaly cases were identified in 23 studies together with the study presented lastly by Singh and Pandey<sup>21</sup> Almost all publications of double RCA anomalies were made in the last 10 years. Most of the 28 cases of double RCA were male. In the majority of cases, double RCA originated from a single ostium. A conus branch arising separately, or a right ventricular branch which is directly originated from right sinus of Valsalva may lead to misdiagnosis of pseudo double RCA. However, a double RCA anomaly originated with separate ostia may not be detected by conventional coronary angiography.<sup>22,23</sup>

In our case, double RCA was originating from a single ostium and both of the RCAs were terminating as PDAs. Therefore, the case was accepted as a real double RCA anomaly. In addition to double RCA anomaly, we detected double fistula from RCA to the right ventricle and from distal LAD to the left ventricle.

Double RCA anomaly is generally considered a being condition. Nonetheless, in recent years, new publications have indicated that double RCA anomaly may be complicated with atherosclerosis.<sup>24</sup> Atherosclerosis was found in nine of the 28 cases described in the literature; eight patients presented with acute coronary syndrome and four with inferior myocardial infarction.<sup>25</sup>

The treatment options for coronary arterial fistulae include surgery or catheter closure. Surgical closure has been reported, either by external ligation of the fistula or by internal patching of the orifice.<sup>26</sup> Risk of surgery was related to cardiopulmonary bypass and median sternotomy.<sup>27</sup> Catheter closure of the fistulae is now considered to be an effective and safe alternative to surgery.<sup>28</sup> The use of implantable coils is currently considered to be the best method, due to improved control and delivery techniques. The risks of transcatheter occlusion is low to date one fatality in an adult patient has been reported.

In our case, patient was discharged because of his small fistulae and  $Q_p/Q_s < 1,3$ . We contem-

plated to closure of fistulae after marked left-to-right shunt is detected during the follow-ups performed in 6-month intervals.

## CONCLUSION

Identification of a coronary artery anomaly by imaging procedures is important because of its impact on the premature cardiac mortality and morbidity. In addition, the possibility of a double RCA arising from separate ostia should always be remembered prior to cardiac interventions. Accurate identification of a coronary artery anomaly may be difficult by conventional angiography and it can be overlooked if clinician does not consider the possibility of such a case.

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