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# Conventional and computed tomography angiography views of a rare type of single coronary artery anomaly: right coronary artery arising from distal left circumflex artery

*Tek koroner arter anomalisinin nadir bir tipinin konvansiyonel ve çok kesitli bilgisayarlı tomografi anjiyografi görüntüleri: Distal circumfleks arterden çıkan sağ koroner arter*

## Introduction

Single coronary artery anomaly (SCA) is defined as the coronary artery arising from a single coronary ostium, supplying the entire heart. Although the incidence of coronary artery anomalies ranges from 0.6% to 1.3% in angiography series, the prevalence of SCA was only found to be 0.02% in the population (1). SCA anomalies are usually benign and asymptomatic; however, serious complications such as sudden cardiac death and myocardial infarction resulting from these anomalies were also reported in the literature.

Right coronary artery (RCA) originating from left coronary sinus or proximal portions of left coronary arteries or left coronary system originating from right coronary sinus constitute the major proportion of SCA anomalies.

Herein, we report a case in which the RCA originates from the distal portions of left circumflex artery as a continuum of it. In addition to conventional angiography images; multi-detector computed tomography (MDCT) was used to confirm the diagnosis and determine the course of the anomalous coronary arteries in this case report.

## Case Report

A 52-year old woman with hypertension and dyslipidemia was admitted to our clinics with class II exertional chest pain according to Canadian Cardiovascular Society classification. After 2 mm horizontal ST depression in the lateral leads with a Duke score of -10 was revealed on stress electrocardiography, coronary angiography was performed. Single coronary artery ostium was detected in which RCA was arising as a continuum of the left circumflex coronary artery (Fig. 1). To confirm this diagnosis and search for a possible cardiac anomaly, which may explain the patient's symptoms, 64-slice MDCT (Aquilion; Toshiba Medical Systems, Tokyo; Japan) was performed thereafter (Fig. 2). With the help of this method, we confirmed the SCA originating from solitary coronary ostium without an additional cardiac anomaly. The RCA was found to be

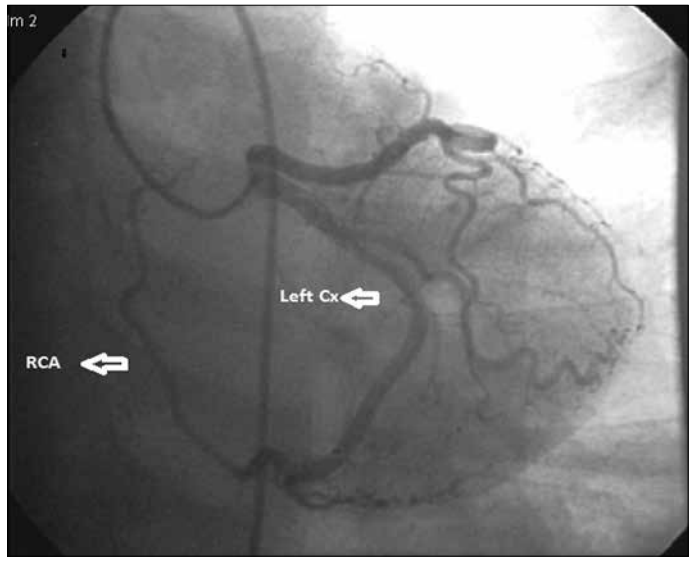


**Figure 1. Conventional angiography image of the single coronary artery**
Cx - circumflex artery, RCA - right coronary artery

**Figure 2. Multidetector computed tomography (64-slice) views of single coronary artery**
Cx - circumflex artery, LAD - left anterior descending artery RCA - right coronary artery

continuous with the distal portion of the left circumflex artery. We decided to continue her medical therapy and added a beta -blocker because we thought that this anomaly might directly induce myocardial ischemia. She had been asymptomatic at her last visit.

## Discussion

Isolated SCA anomaly is one of the rarest coronary anomalies and constitutes 2-4% of all the coronary artery anomalies. SCA has been reported to be seen in 0.024% to 0.066% of the patients who undergo diagnostic coronary angiography (1-3). Our case is a very rare type of SCA anomaly and according to the Shirani et al. (4) classification, it can be categorized into the IA group which means that a solitary ostium in the left aortic sinus (I) is unassociated with an aberrant-coursing coronary artery (anatomic SCA) (A). This type has been reported in a few numbers in the literature (5, 6).

SCA anomalies are usually found incidentally during coronary angiography. Sudden death and myocardial infarction after exercise have been reported in patients whose left main or right coronary artery goes between main pulmonary artery and aorta (7). Shirani et al. (4) demonstrated that 15% of patients with SCA might have coronary ischemia due to the relation of coronary arteries with aorta or pulmonary artery. Thus, a coronary anomaly may itself cause myocardial ischemia without contribution of significant coronary stenosis.

Myocardial ischemia has been reported in 2 cases whose RCA originates from the left anterior descending or circumflex artery (8). In these cases, thinning of coronary arteries especially RCA was supposed to be responsible for cardiac ischemia. Herein, we presented the most benign type of SCA anomaly (2, 6) which was confirmed by MDCT. In our case, atherosclerosis, presence of which is an important prognostic factor in this type of SCA anomaly (2), was not present in the coronary arteries. We thought that ischemia caused by the SCA anomaly due to the thinning of RCA, was relieved by adding a beta- blocker.

## Conclusion

This is the first case report on both conventional angiography and the MDCT images of a RCA arising from distal left circumflex artery.

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# Char syndrome, a familial form of patent ductus arteriosus, with a new finding: hyperplasia of the 3rd finger

## *Ailesel patent duktus arteriyozus: Char sendromu ve yeni bir bulgusu; 3. parmak hipoplazisi*

## Introduction

Char syndrome is an autosomal dominant disorder characterized by patent ductus arteriosus (PDA), facial dysmorphism and abnormalities of the fifth finger of the hand (1). The prevalence of Char syndrome has not been determined but is believed to be quite low.

This report describes a Turkish family including five individuals affected by this disorder with an R236C mutation in the gene encoding the neural-crest-related transcription factor AP-2b. Affected family members had the typical facial, hand and foot anomalies and additionally presented case has rarely reported polythelia and non reported hypoplasia of the 3rd finger.

## Case Report

A 15-day-old girl was referred because of a cardiac murmur. Consanguinity between the parents was denied. The respiratory and heart rates were 80/min and 160/min respectively, The patient had a flat midface, widely set eyes, mild ptosis, short philtrum and a triangular mouth; polythelia, foot and hand anomalies with clinodactyly were also noted (Fig. 1). Echocardiography revealed a large duct (6.5 mm) with unrestrictive ductal flow and predominantly left-to-right shunting, leading to left heart volume overload. The patient had an uneventful follow-up after surgical ligation and was discharged on the postnatal 45th day. The family history was suggestive for the presence of Char syndrome. His father, paternal uncle and a cousin were operated on for PDA. Similar phenotypic features and variable hand-foot anomalies were seen in them (Fig. 2). Additionally his paternal grandmother has typical facial dysmorphism, a small PDA, and polythelia. The pedigree is shown in Figure 1. Hypoplasia of the 3rd finger as a new finding in this syndrome was found in the proband and his father. Developmental, visual and hearing disorders were not detected in any members.

Genetic analysis of the *TFAP2B* coding exons and their flanking exons was performed as previously described (2). Analysis of the proband's genomic DNA revealed a coding region alteration in exon 4, a C-to-T transition at nucleotide 706 of the *TFAP2B* cDNA, which was present in heterozygosity. This sequence change predicted a substitu-