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# Isolated Single Coronary Artery
## — A Series of 10 Cases —

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**Background** Isolated single coronary artery (SCA) is an extremely rare congenital coronary anomaly. Some subgroups of SCA can lead to angina pectoris, acute myocardial infarction or even sudden death in the absence of atherosclerosis. Young patients, especially, have the risk of serious clinical events, but middle-aged-to elderly patients have a variable clinical course.

**Methods and Results** The aim of this study was to present the clinical and angiographic properties, relatively long-term follow-up (54±14 months) and management of adult patients (mean age 57±12 years) with SCA. The records of 70,850 patients undergoing coronary angiography between 1999 and 2005 were reviewed. Ten patients (0.024%) were found to have SCA, originating from the left sinus of Valsalva in 3 (30%) patients and from the right sinus of Valsalva in 7 patients (70%). No atherosclerotic involvement was seen in 7 (70%) patients. One patient was also treated by stent implantation to the SCA. Other patients were followed medically. All patients have been followed uneventfully.

**Conclusion** Medical treatment is usually adequate for middle-aged to elderly patients with SCA in the absence of ischemia and/or acute coronary syndrome. (*Circ J* 2008; **72:** 1254 – 1258)

**Key Words:** Angiography; Anomaly; Coronary artery anomalies; Imaging; Single coronary artery

Single coronary artery (SCA) is a rare congenital anomaly whereby only 1 coronary artery arises from the aortic trunk from a single coronary ostium, supplying the entire heart.[1] The incidence of isolated SCA is only 0.0024–0.066% in the general population undergoing coronary angiography (CAG).[2–4] SCA may be associated with other severe congenital cardiac malformations (persistent truncus arteriosus, tetralogy of Fallot, pulmonary atresia).[5,6] The first case of SCA was reported by Thebesius in 1716 and Halperin et al made the first antemortem diagnosis by means of CAG in 1967.[4] The clinical significance of SCA is unknown and although it is generally considered benign, some authors have studied sudden death associated with isolated congenital coronary anomalies in an autopsy population and observed that sudden death occurred in patients with SCA.[7] There is a lack of data about the angiographic and clinical characteristics and long-term follow-up of these patients, so in the present study, we describe the angiographic variations, relatively long-term follow-up data (50±14 months) and clinical findings of 10 SCA cases.

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## Methods

Between 1999 and 2005, we surveyed the records of 70,850 adult patients who had undergone diagnostic CAG at the Medical Center of Sani Konukoglu, University of Harran, Technical University of Black Sea, University of 100th years and Cardiology Hospital of Hanefi Oksuz.[8]

The CAG findings and clinical data were analyzed. Charts were reviewed for patients' demographics, clinical presentations and findings at cardiac catheterization. CAG was performed either by the Judkins femoral method or the Sones brachial method in each center. The course of the anomalous coronary artery was determined from different CAG views (ie, left anterior oblique, right anterior oblique and lateral views).

The records of each patient were examined by at least 2 experienced angiographers who grouped the anomalies according to the very useful angiographic classification developed by Lipton et al[2] and further modified by Yamanaka and Hobbs.[3] Based on those classifications, the first division was made between the 'R' right-type and 'L' left-type according to the site of origin of SCA (ie, in the right or left sinus of Valsalva). Each case was then designated as belonging to group I, II or III depending on the anatomical course of the artery. Group I had an anatomical course of either a right or left coronary arteries (RCA or LCA); for example, when the artery originates from the right sinus of Valsalva, it follows the course of a normal RCA, giving off the posterior descending artery in normal fashion near the crux and then continuing in the left atrioventricular groove to give off the posterolateral left ventricular branches. Group II anomalies arise from the proximal part of the normal RCA or LCA, and cross the base of the heart before assuming the normal position of the inherent coronary

**Table 1 Classification of Angiographic Types of Single Coronary Artery (Modified Lipton's Classification[2])**

| Code | 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 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 sinus of Valsalva (R-III)* |
| *Course of the transverse trunk* | |
| *A* | *Anterior to great vessels* |
| *B* | *Between 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 anterior descending coronary artery; LCX, left circumflex coronary artery.*

**Table 2 Properties of the Single Coronary Artery Anomalies**

| Case no. | Sex | Age (years) | Symptom | Rest/stress ECG | Modified Lipton's classification | Atherosclerotic involvement | Treatment | Follow-up (months) |
|---|---|---|---|---|---|---|---|---|
| 1 | M | 35 | SAP | ST-T wave changes | Left, Type II-A (Fig 1) | – | Medical | 34 |
| 2 | M | 66 | UAP | ST-T wave changes | Left, Type I (Fig 2) | LAD | Medical | 64 |
| 3 | M | 55 | SAP | | Right, Type I (Fig 3) | – | Medical | 36 |
| 4 | F | 54 | SAP | | Right, Type II-A | – | Medical | 62 |
| 5 | M | 78 | SAP | | Right, Type II-A (Fig 4) | – | Medical | 50 |
| 6 | F | 57 | SAP, Palpitation | ST-T wave changes | Left, Type I | – | Medical | 66 |
| 7 | M | 70 | SAP, Dyspnea on exercise | Treadmill (+) | Right, Type II-B* | CX, RCA | PCI | 68 |
| 8 | F | 48 | SAP, Palpitation | ST-T wave changes | Right, Type III (Fig 5) | – | Medical | 48 |
| 9 | M | 57 | SAP | | Right, Type III (Fig 6) | – | Medical | 33 |
| 10 | F | 47 | SAP | ST-T wave changes | Right, Type III | RCA | Medical | 39 |

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*SAP, stable angina pectoris; UAP, unstable angina pectoris; CX, circumflex coronary artery; PCI, percutaneous coronary intervention. Other abbreviations see in Table 1.*

artery. The subgroup designation of Group II refers to the relationship between the anomalous coronary artery and the aorta and pulmonary artery. The letters 'A', 'B' and 'P' refer to 'anterior', 'between', and 'posterior' patterns. Group III describes the anomaly where the left anterior descending (LAD) coronary artery and left circumflex (LCX) coronary artery arise separately from the proximal part of the normal RCA (Table 1).

A vessel was considered to have significant stenosis if its diameter, by visual estimation, was narrowed by >70% (for left main >50%), with respect to the adjacent pre-stenotic segment. The criteria used for selection of the interventional procedure or medical treatment were based on the presence of symptoms and CAG characteristics, as well as associated cardiac conditions. The patients were given β-adrenergic antagonists, nitrates, calcium-channel blockers, statins or antiplatelets as their medical treatment.

The patients were telephoned every 6 months during the follow-up period and their follow-up data were obtained by questionaire. Follow-up data included symptom status and information about mortality.

### *Statistical Analysis*

Data are presented as mean±SD, percentage or range.

## Results

### *Patients and Their Clinical Presentations*

In this study of 70,850 patients who had diagnostic cardiac catheterization, 10 (prevalence of 0.014%) had SCA (clinical and angiographic findings are summarized in Table 2). Their mean age was 57±12 years (range 35–78), 6 were males (60%), with a mean age of 60±14 years and 4 (40%) were females, with a mean age of 52±5 years.

Of the 10 patients, 7 did not have atherosclerotic coronary artery disease (70%) and all had stable angina pectoris. Two patients had both palpitations and stable angina pectoris. In 3 of the 7 patients, ECG revealed nonspecific ST-T changes (30%). ECGs of the other 4 patients were normal.

Of the 3 patients with significant coronary artery disease (30%), 1 was followed in hospital for the diagnosis of unstable angina pectoris and her ECG showed dynamic ST-T changes during the hospitalization. The second patient was suffering from angina and dyspnea induced by exercise. Although the resting ECG showed normal sinus rythm, the exercise ECG was positive. The third patient also had stable angina pectoris and a normal resting ECG.

### *CAG Characteristics*

In 3 of the 10 patients, the SCA originated from the left sinus of Valsalva (30%) and in 1 of these patients, the SCA gave off the LAD and circumflex coronary artery (CX) in the usual pattern. The RCA, as the branch of the LAD, was going ahead of the aorta and was nondominant coronary artery (Left, Type II-A) (Fig 1). In the other 2 patients (Cases 2 and 6, Table 2), the SCA gave off the anterior descending branch in the usual pattern and then continued in the atrioventricular groove as the LCX branch. It traveled beyond the crux into the right atrioventriculer groove where it provides branches to the right ventricle and atrium (Left,

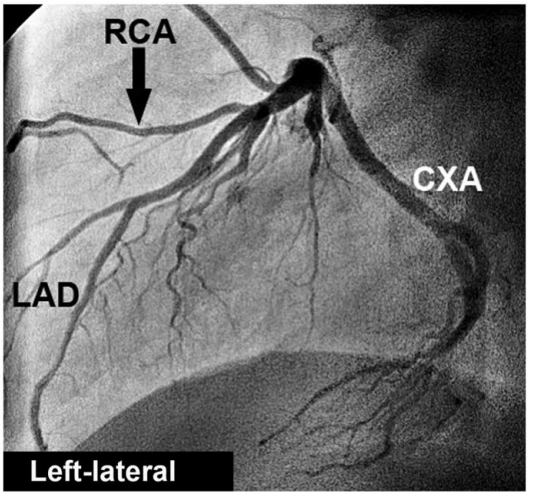



Fig 1. Left lateral view of anomalous single coronary artery arising from the left sinus of Valsalva (Left, Type II-A). CXA, circumflex coronary artery; LAD, left anterior descending artery; RCA, right coronary artery.

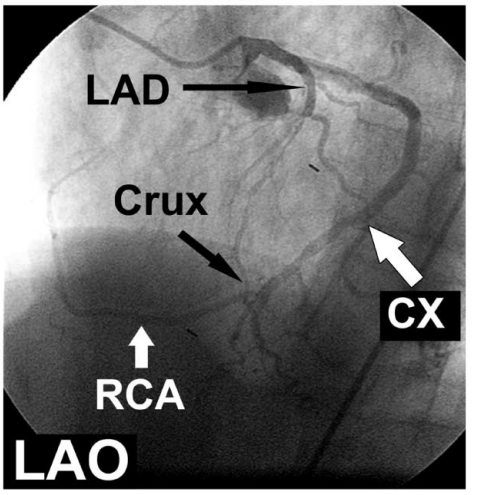



Fig 2. Representative anomalous single coronary artery from the left sinus Valsalva (Left, Type I). The LAD shows significant atherosclerotic involvement. LAO, left anterior oblique; LAD, left anterior descending coronary artery; RCA, right coronary artery; CX, circumflex coronary artery.

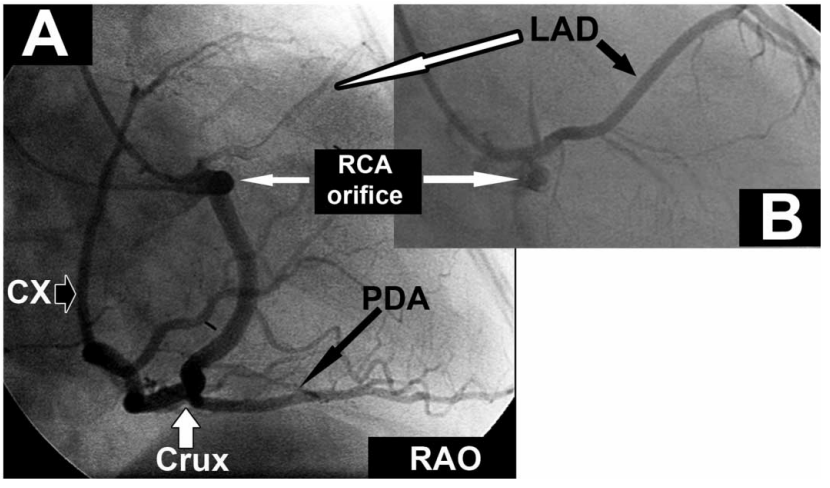



Fig 3. RAO view of anomalous single coronary artery arising from the right sinus of Valsalva while giving off a thin LAD branch at the beginning (Right, Type I). RAO, right anterior oblique; RCA, right coronary artery; LAD, left anterior descending artery; CX, circumflex coronary artery; PDA, posterior descending artery.

Type I). There was a serious atherosclerotic lesion in the LAD of 1 of these patients (Fig 2).

In 7 patients the SCA originated from the right sinus of Valsalva (70%). In 1 of them (Case 3, Table 2), the SCA began as the RCA that continued until the crux where it gave off a thin LAD branch at the beginning. Beyond the crux it travelled as the dominant CX, providing posterior descending branches (Right, Type I). There was not a coronary atherosclerotic lesion in the coronary tree (Figs 3A,B). In 2 patients (Cases 4 and 5, Table 2), the SCA began from the right sinus of Valsalva, providing the main coronary branch that supplied the LAD and CX that travelled in front of the aorta and gave off the dominant RCA branch (Right, Type II-A). There was not a coronary atherosclerotic lesion in the coronary tree (Fig 4). In a previously published case (Case 7, Table 2), the SCA arose from the right sinus of Valsalva. The left main coronary artery branch, originating from the SCA, was travelling between the aorta and pulmonary artery and then provided branches as the LAD and CX. In that case, although the RCA went its normal course, the sinus node branch originated from the CX artery. Moreover, there was an atherosclerotic lesion in the RCA and CX, and the patient showed moderate mitral valve regurgitation, for which percutaneous coronary angioplasty was performed to the CX branch (Right, Type II-B)[9]. In the other 3 patients, the SCA originating from the right sinus of Valsalva was providing the LAD, CX and RCA branches (Cases 8–10, Table 2) (Right, Type III) (Fig 5). In those patients, the LAD and CX branches were going ahead of the aorta. In 1 case, only 1 diagonal branch originating from the LSV was seen separately (Fig 6). To our knowledge, this angiographic combination is reported here for the first time in the literature.

*Treatment and Survival*

Percutaneous coronary intervention was performed in 1 patient only. Other patients were followed medically. As medical treatments, β-blockers, nitrates, calcium-channel blockers, statins or antiplatelets in particular were appropriately selected for the patients. Our approach included recommendations related to avoiding heavy exercise. The symptoms of the patients improved following medical or revascularization therapy.

The follow-up period ranged from 33 to 68 months

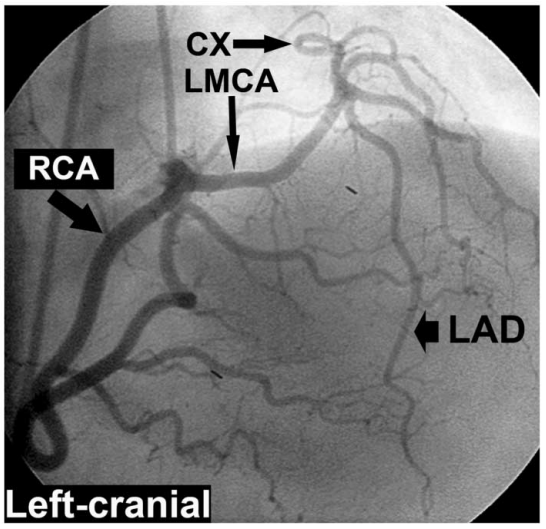



Fig 4. Left-cranial view of anomalous single coronary artery arising from the right sinus of Valsalva, providing the main coronary branch, which supplies the LAD and CX that traveled in front of the aorta (Right, Type II-A). LMCA, left main coronary artery; RCA, right coronary artery; LAD, left anterior descending coronary artery; CX, circumflex coronary artery.

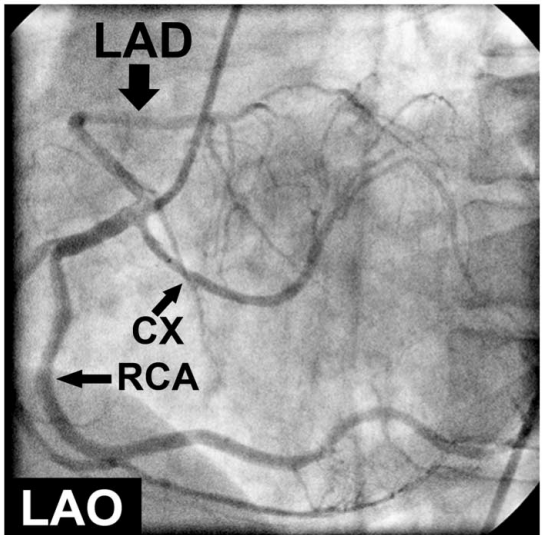



Fig 5. LAO view of anomalous single coronary artery arising from the right sinus of Valsalva (Right, Type III). LAO, left anterior oblique; LAD, left anterior descending coronary artery; RCA, right coronary artery; CX, circumflex coronary artery.

(50±14 months). During the follow up period, there were no deaths directly attrubutable to the SCA anomaly. The medical treatment of the 10 patients was regular and they are free of symptoms.

## Discussion

Coronary artery anomalies are not seen frequently during routine cardiac catheterization and the incidence of anomalous coronary artery is reported to range from 0.2% to 1% in patients undergoing CAG and 0.3% in autopsies[3,8] A SCA ostium, in the absence of other cardiac disease, is an extremely rare coronary artery anomaly. SCA constitutes approximally 2–4% of all coronary anomalies, and isolated

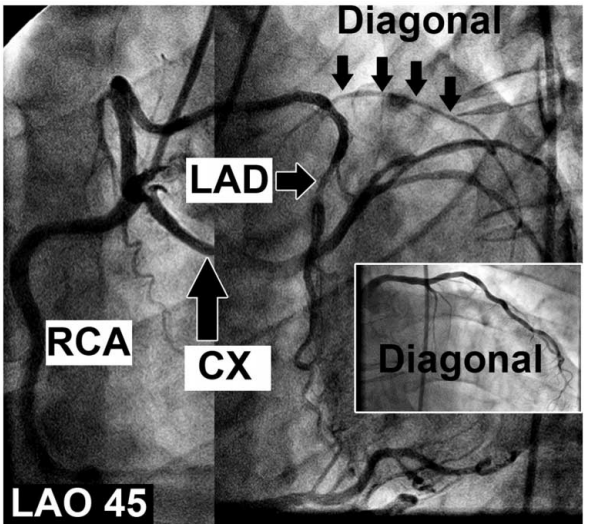



Fig 6. Left anterior oblique view of anomalous single coronary artery arising from the right sinus of Valsalva (Right, Type III). Only 1 diagonal branch originates from the left sinus of Valsalva. LAO 45, left anterior oblique 45°; RCA, right coronary artery; LAD, left anterior descending coronary artery; CX, circumflex coronary artery.

SCA occurs in approximately 0.024–0.066% of the population[2–4] In the present study, review of 70,850 records of adult patients who had undergone diagnostic CAG between 1999 and 2005, found an SCA prevalence of 0.014%.

These anomalies are commonly an incidental finding during CAG. The prognostic significance varies, with the majority of patients being asymptomatic. However, 15% of patients may have myocardial ischemia directly caused by the abnormal anatomy of the arteries and not by coronary artery disease[10] Myocardial perfusion can be affected, ranging from exertional angina to sudden death in the very uncommon subtypes of these anomalies[2,11] Ischemia is the consequence of anatomical malformations, including the acute angle take-off of the anomalous vessel, with a narrowed slit-like orifice that collapses in a valve-like manner, thereby limiting the blood flow. Other anatomical features responsible for ischemia are the proximal intramural course of the anomalous vessel, which is squeezed within the aortic wall, and the compression of the anomalous vessel along its course between the aorta and the pulmonary artery, particularly during exercise[12,13]

Extremely rare Group I (R-I, L-I type) anomalies generally have a benign clinical course[14] The R II-III type and L II-III type anomalies having a coronary artery branch following a dangerous course, which may predispose to clinical effects. Basso et al claim that of the patients with SCA, most (59%) die under 30 years of age, usually during or shortly after vigorous exertion[13–15] In addition, coronary artery anomalies are the second cause of sudden death in young athletes[16] Accordingly, the low prevalence in our study may be related to the composition of the patient population, being mostly middle-aged-to elderly, and the loss of anomaly cases under 30 years because of cardiac causes. That result suggest that the angiographic features of the patients in our study were generally benign.

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 bicus-

pid aortic valve[1,17] Therefore, an accompanying coronary artery anomaly must be considered as likely in such cases that were stabilized as congenital heart disease.

Another characteristic of clinical importance is encountered during surgery on patients with a congenital anomaly. Complications may arise if the operator unwittingly incises the anomalous vessel. The surgical approach may involve 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 with good long-term results.

CAG is still the gold standard, and required to accurately diagnose and evaluate coronary anomalies. It gives a 2-dimensional view of the coronary vasculature. High image resolution, complex 3-dimensional view and non-invasive nature have made multislice computed tomography and magnetic resonance imaging angiography very useful in the exclusion of atheromatous coronary artery disease and determination of the coronary anatomy[18,19] If there is any suspicion of a coronary anomaly in young cases (under 30 years) of congenital heart disease, these noninvasive investigations should be used. Also, ECG-gated single-photon emission computed tomography enables useful noninvasive evaluation of both myocardial blood flow and cardiac function, and thus it may be suitable to show the ischemia caused by the coronary anomaly[20–22]

Unfortunately, there still is not a consensus on the management of SCA. Group I (R-I, L-I type) anomalies are generally treated medically, but surgery must be considered for patients under 30 years, especially those with the R II-III type and L II-III type anomalous SCA, if there is documented ischemia without arterosclerotic involvement. It is unclear whether surgical intervention currently improves the long-term outcome. On the other hand, long-term data about medical management are also lacking. Without this crucial clinical data, it is undoubtedly difficult to set guidelines/recommendations for surgical correction of this condition. In the present study, we preferred medical treatment in the absence of associated severe coronary artery disease and/or overt ishemia.

*Study Limitations*

As a result of patients being discovered retrospectively and the study being multicentered, the clinical data and laboratory data of patients at the time of admission was not detailed. Another limitation of this study was the relatively small sample size.

## Conclusions

Coronary artery anomalies are encountered more often in the daily practice of high-volume cardiac centers. Cardiac surgeons and interventional cardiologists should be aware of these pathologies because recognition of them on CAG is mandatory for the prescription of appropriate medical or surgical therapy. However, there are insufficient data about the optimal management of this specific family of coronary artery anomalies. According to the results of our long-term follow-up study (50±14 months) we can say that medical treatment is usually adequate for SCA in middle–aged-to elderly patients in the absence of ischemia and/or acute coronary syndrome. But we still need more data about the natural history, clinical findings and angiographic properties of this rarely seen coronary anomaly.

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