

# Single coronary artery incidence in 215,140 patients undergoing coronary angiography

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**Background:** The aim of our study is to determine the incidence of single coronary artery (SCA). SCA is a rarely seen coronary anomaly in which the right coronary artery and the left main coronary artery arise from single aortic sinus. Although SCA has a benign course in most cases and its clinical significance is unknown, in some autopsy studies it was shown to be related to sudden cardiac death.

**Materials and methods:** SCA patients detected among 215,140 coronary angiographies (CAG) performed between 1998 and 2013 in SANKO Hospital were included in our study. The classification of CAG was made according to the two different classifications defined by Smith and Lipton and colleagues.

**Results:** A total number of 215,140 patients who underwent routine CAG were included in the study, and SCA was detected in 67 (0.031%) patients. There were 6 (9%) type R-I, 23 (34%) type R-II, 10 (15%) type R-III, 16 (24%) type L-I and 12 (18%) type L-II patients according to the angiographic classification.

**Conclusions:** SCA is rarely seen during routine cardiac catheterisation and its incidence is 0.014–0.066% in angiographic series. In our study, the incidence was shown to be similar to the previous studies. (Folia Morphol 2014; 73, 4: 469–474)

**Key words:** coronary vessel anomalies, coronary angiography, incidence

## INTRODUCTION

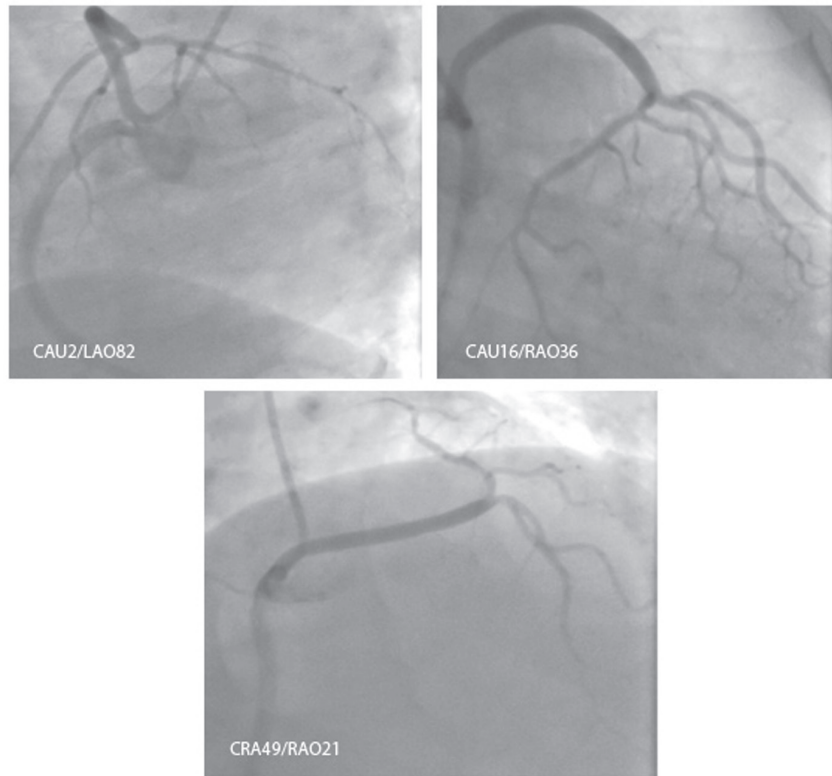
Single coronary artery (SCA) is a rarely seen coronary anomaly in which right coronary artery (RCA) and left main coronary artery (LMCA) arise from single aortic sinus (Figs. 1, 2). It is detected usually during routine coronary angiography (CAG) or found incidentally in postmortem examination [18]. It has an incidence of 0.014–0.066% in general population [6, 10]. Congenital heart diseases like tetralogy of Fallot, transposition of great arteries, patent ductus arteriosus, coronary arteriovenous fistulae and bicuspid aorta accompany SCA in 40% of the cases [4, 13]. Kervancioğlu et al. [9] reported that SCA had a rate of 3.5% among 607 Fallot patients. Thebesius reported the first SCA case in 1,716, and the first

antemortem diagnosis was made by Halperin et al. angiographically in 1967 [6].

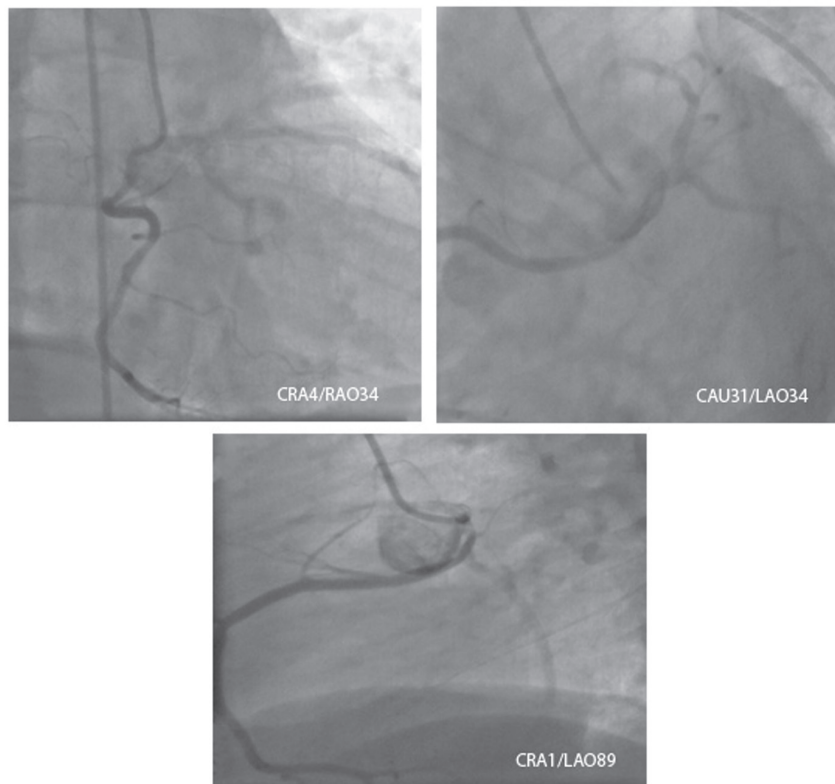
Although SCA has a benign course in most cases and its clinical significance is unknown, in some autopsy studies it was shown to be related to sudden cardiac death (SCD) [7]. Herein we evaluated retrospectively the incidence and angiographic characteristics of SCA patients diagnosed in the past 15 years.

## MATERIALS AND METHODS

SCA patients detected among 215,140 CAGs performed between 1998 and 2013 in SANKO Hospital were included in our study. Patients with SCA were selected retrospectively from coronary anomaly files created during that period, and the incidence and



**Figure 1.** Angiographic view of single coronary artery originating from the right sinus of Valsalva.



**Figure 2.** Angiographic view of single coronary artery originating from the left sinus of Valsalva.

clinical features of SCA patients were determined. Electrocardiographies (ECGs), treadmill exercise tests and coronary artery disease (CAD) risk factors were obtained from patient files. Ischaemic ST-T changes in ECG were examined. Treadmill exercise test was performed according to the Bruce protocol and presence of 1 mm or greater, horizontal or downsloping ST segment depression in at least 2 leads and/or 30 mm Hg decrease in systolic blood pressure and/or ventricular arrhythmias and/or typical angina were accepted as a positive test.

CAG was performed by femoral (Judkins), brachial (Sones) or transradial optitorque method with Siemens Axiom Angiography device. The route of anomalous coronary artery was evaluated in at least 4 different anatomical positions (left lateral, antero-posterior caudal and cranial or left oblique).

The patients were allocated into four groups according to their age: 20–29, 30–39, 40–49, and 50 years and above.

The classification of CAG was made according to the two different classifications defined by Smith and Lipton and colleagues. The Smith's system consists of three groups according to the course of the coronary arteries involved: In group I, a SCA courses as the RCA, circumflex (LCX) or as the left anterior descending artery (LAD), or a single LMCA is divided into 2 branches as the LAD and LCX, and RCA is formed by the extension of the LCX traversing the base of the heart. In group II, the main single artery either gives off right and LMCA arteries, or RCA, LAD, and LCX separately, which then follows their original paths. In group III, the single coronary artery shows an atypical branching that leads to significant differences in the original routes of the 3 main coronary arteries. Lipton and colleagues classified the coronary anomalies as R or L according to the origination of abnormal coronary artery either from right or left coronary sinus (Figs. 1, 2).

A stenosis of a vessel was considered significant if its diameter was narrowed by > 60% (for left main > 50%) compared to the adjacent normal segment. The choice of interventional procedure or medical treatment was made by visual estimation and based on the presence of symptoms, CAG characteristics and associated cardiac conditions. The patients were given beta-adrenergic antagonists, nitrates, calcium-channel blockers, statins or antiplatelets for medical treatment.

The statistical analysis of the study was performed by SPSS 14 statistics (SPSS for Windows 14, Inc., Chi-

**Table 1.** Patient characteristics with single coronary artery

Age [year]	48 ± 11 (29–75)
Gender (male/female)	40/27
Hypertension	22 (33%)
Diabetes mellitus	14 (21%)
Hyperlipidaemia	14 (21%)
Smokers	30 (45%)
Myocardial infarction	3 (4.5%)

**Table 2.** Single coronary artery types

R-I	6 (9%)
R-II	23 (34%)
R-III	10 (15%)
L-I	16 (24%)
L-II	12 (18%)

cago, IL, USA) programme. The data were presented as percentages and mean ± standard deviation.

## RESULTS

A total number of 215,140 patients who underwent routine CAG were included in the study and SCA was detected in 67 (0.031%) patients. Forty (60%) of the patients were males and 27 (40%) of them were females. The mean age was 48 ± 11 years (minimum 29, maximum 75); 22 (33%) patients had hypertension, 14 (21%) diabetes mellitus, 14 (21%) hyperlipidaemia and 30 (45%) were smokers (Table 1). There were 6 (9%) type R-I, 23 (34%) type R-II, 10 (15%) type R-III, 16 (24%) type L-I and 12 (18%) type L-II patients according to the angiographic classification (Table 2).

We detected 1 patient in 20–29 group, 15 patients in 30–39 group, 24 patients in 40–49 group and 27 patients in 50 and above group.

Twenty-six (38%) patients had pathologic ST-T changes on ECG. Treadmill exercise test was performed in 56 patients and 34 (61%) of 56 patients had positive results. One patient experienced ventricular tachycardia during exercise test and we made electrical cardioversion. In the coronary angiography of this patient, we detected type R-II coronary anomaly and recommended surgery, but the patient refused surgical treatment.

Thirty-nine (58%) patients had experienced exertional angina, 6 (9%) patients had exertional dys-

pnoea, 3 (4%) had syncope, 4 (6%) had palpitations, 6 (9%) had exertional angina and palpitations and 6 (9%) had exertional angina and syncope. Three of the patients were admitted with myocardial infarction (MI) (Table 1). One of these 3 patients was admitted with acute inferior MI, and an R-III type anomaly was detected during CAG. Percutaneous coronary intervention (PCI) was performed for totally occluded RCA. In 1 of 2 remaining patients hospitalised with the diagnosis of non-ST-segment elevation MI, we detected R-II type anomaly and implanted a stent in LCX artery. The second patient had R-II type anomaly, and coronary artery bypass graft (CABG) operation was performed. In 8 (1 patient had type L-I, 2 patients type R-II, 1 patient type R-I, 3 patients type R-III, 1 patient type L-II) of 64 individuals, excluding 3 acute coronary syndrome patients, significant CAD was detected and 3 of them underwent PCI and 5 patients were treated by CABG. Thirty-nine of the other 56 patients had normal coronary arteries and 17 had non-critical atherosclerotic plaques. Corrective surgery was recommended for 10 (3 of them had type R-II, 2 type R-I, 3 type R-III, 2 type L-II) of these 56 patients because of severe exertional dyspnoea and positive treadmill test, however 5 of them refused surgical therapy. Corrective surgery was performed in 2 of 5 patients who accepted surgical treatment and CABG surgery was done in 3 remaining patients. Medical treatment was started in those patients who refused surgery. Beta-blockers, calcium channel blockers, statins and antiplatelet agents were initiated as medical therapy. The patients were suggested to avoid strenuous exercise.

## DISCUSSION

Coronary artery origin anomalies are rarely seen during routine cardiac catheterisation and the incidence is 0.2–1.3% in angiographic series and 0.3% in autopsy series [23, 25]. Most of these anomalies are clinically non-significant, however some anomalies can manifest as angina, dyspnoea, syncope, acute MI and SCD [15]. Their clinical significance is unclear, since they are rare coronary anomalies and there is limited number of patients in previous studies and the follow up periods are insufficient.

SCA constitutes 2–4% of all coronary anomalies and its incidence in population is 0.014–0.066% [6, 10, 21]. In a retrospective analysis of Tuncer et al. [21] with 70,850 patients, its incidence was 0.014%. In this analysis, they reported 2 patients with type L-I,

1 patient with type L-II, 1 patient with type R-I, 3 patients with type R-II and 3 patients with type R-III [21]. In the analysis of Yamanaka et al. [23] with 126,595 patients, the incidence of SCA was found to be 0.045%. In this analysis, there were 20 patients with type L-I, 11 patients with type L-II, 1 patient with type R-I, 19 patients with type R-II and 5 patients with type R-III [23]. In a retrospective report of with 50,000 patients, Desmet et al. [6] detected SCA in 33 patients and the incidence was reported to be 0.66%. In our data set including 215,140 CAGs from 15 years, there are 67 patients with SCA and the incidence is 0.31%. The highest SCA rate was detected in 40–49 age group.

Ischaemia induced by tachycardia and increased pulsatility and stroke volume during exercise can explain the association between SCA and sudden death during heavy exercise [22]. Adults and older patients can be admitted due to new onset angina, palpitations, ECG changes, dyspnoea, syncope and SCD triggered especially by exercise [23].

In cases of abnormally originating LAD from RCA, LAD may run between pulmonary artery and aorta, over septum, anteriorly, retroaortic or posterior to the aorta [12]. Septal path is usually benign and sudden death was not reported so far [12]. In case of anomalous vessel running between aorta and pulmonary artery, lateral compression and kinking of anomalous artery between aorta and pulmonary artery lead to exertional dyspnoea and angina or trigger myocardial ischaemia which causes serious malign ventricular arrhythmias and SCD [2, 19, 20]. This condition is related to SCD and worse prognosis [7]. This compression can be seen if the retroaortic coronary artery does not lie within the aortic wall, and this kind of anomaly has been reported only in 1 case [13]. Nevertheless, ischaemia can be seen if an anomalous coronary artery does not run between the great vessels [16].

In 15% of patients, myocardial ischaemia can be detected in the absence of CAD due to abnormal coronary anatomy [18]. Ischaemia is the result 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 limiting the blood flow [2, 5]. We performed exercise test in 56 patients and 34 (61%) of them had positive test results.

Angiographically SCA is classified according to the abnormal origin, course, branching pattern and termination of the vessel [3]. R-I is the most common

form and usually has a benign clinical course [17]. L1 is also generally benign. Although previous reports showed that L-I is rare, among 126,595 angiographies of Yamaka et al. [23], 20 of 56 SCA cases had L-I type anomaly. In our study, we detected 16 (24%) L-I anomalies among 67 SCA cases. R-II, R-III, L-II, L-III types are more commonly seen anomalies and they can have more severe clinical courses [5, 14]. Basso et al. [5] claimed that more than half of the patients with SCA die before 30 years of age during or soon after strenuous exercise [14].

Coronary artery anomalies are the second most common cause of the SCD in young athletes [11]. In an autopsy examination of 27 athletes with SCD, Basso et al. [5] reported 23 LMCA originated from right aortic sinus and 4 RCA originated from left aortic sinus. Because of this reason, heavy exercise seems to be the most significant trigger of death in patients with SCA [5]. There was not any cardiovascular symptoms or tests performed before in 15 of these patients, however, 10 of 12 patients had experienced symptoms like palpitations, dyspnoea, angina and syncope previously [5].

CAG is the gold standard technique in the accurate assessment of the anatomy. Additionally, multislice computed tomography and magnetic resonance imaging can be useful in the evaluation of the coronary anatomy because of their higher image resolution, complex 3-dimensional imaging and non-invasiveness. ECG-gated single-photon emission computed tomography as a non-invasive procedure provides concomitant evaluation of myocardial blood flow and cardiac functions and it is a useful tool in the diagnosis of ischaemia due to coronary anomaly [8, 24]. Non-invasive tests are recommended preferably in young individuals having congenital heart disease if there is suspicion of coronary anomalies [8, 24].

Surgical management consists of re-implantation of abnormal vessel into the correct coronary sinus or CABG alone in order to provide distal flow in normal coronary artery [1]. It should be kept in mind that unless SCA is not recognised during the operation of accompanying congenital cardiac diseases, this situation can result in resection of abnormal vessel leading to undesired complications [1]. Additionally, percutaneous interventions in these patients can cause ostial dissection and thrombosis leading to catastrophic events [1].

The management strategies in SCA is not clear because of the limited number of the previous cases.

Medical follow-up is recommended for group I because of its benign character, however surgical correction is considered if ischaemia is documented in the absence of atherosclerosis especially in patients under 30 years with type II and III anomalies. However, the long term outcomes of medical therapy or surgery are insufficient. We preferred surgery in patients with significant CAD or documented ischaemia, and in patients without these conditions we chose medical follow-up. We performed PCI in 2 patients with MI and 3 patients with stable angina pectoris (SAP). CABG was recommended for 1 patient after MI and 5 patients with SAP who had multiple vessel coronary disease, Corrective surgery was done in 2 of 4 patients without atherosclerosis, and CABG was performed in 2 remaining patients.

#### Limitations of the study

Our study was conducted by selection of patients from coronary anomaly files collected during 15-year period in our clinic. Since the communication or address data of some patients was wrong or deficient and a significant number of the patients did not return for control visits, we could not generate any follow-up data.

## CONCLUSIONS

In our retrospective study, we evaluated and summarised the incidence and angiographic characteristics of SCA diagnosed in the last 15 years. Our patient group consisted of the patients detected during routine CAG and most of the them were middle aged individuals. In our study, the incidence was shown to be similar to the previous studies.

## REFERENCES

1. Akcay A, Tuncer C, Batyraliev T, Gokce M, Eryonucu B, Koroglu S, Yilmaz R (2008) Isolated single coronary artery: a series of 10 cases. *Circ J*, 72: 1254–1258.
2. Angelini P (2002) Coronary artery anomalies-current clinical issues: definitions, classification, incidence, clinical relevance, and treatment guidelines. *Tex Heart Inst J*, 29: 271–278.
3. Angelini P (2007) Coronary artery anomalies an entity in search of an identity. *Circulation*, 115: 1296–1305.
4. Antonellis J, Rabouni A, Kostopoulos K, Margaris N, Kranidis A, Salahas A, Ifantis G, Koroxenidis G (1996) Single coronary artery from the right sinus of Valsalva, associated with absence of left anterior descending and an ostium-secundum-type atrial septal defect: a rare combination: a case report. *Angiology*, 47: 621–625.
5. Basso C, Maron BJ, Corrado D, Thiene G (2000) Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden

- death in young competitive athletes. *J Am Coll Cardiol*, 35: 1493–1501.
6. Desmet W, Vanhaecke J, Vrolix M, Van de Werf F, Piesens J, de Geest H (1992) Isolated single coronary artery: a review of 50000 consecutive coronary angiographies. *Eur Heart J*, 13: 39–47.
  7. Frescura C, Basso C, Thiene G, Corrado D, Pennelli T, Angelini A, Daliento L (1998) Anomalous origin of coronary arteries and risk of sudden death: a study based on an autopsy population of congenital heart disease. *Hum Pathol*, 9: 689–695.
  8. Iio K, Nandate H, Nakamura T, Nakashima Y, Kuroiwa A (1989) The study of a case of single coronary artery using stress 201-thallium single photon emission computed tomogram. *J UOEH*, 11: 55–62.
  9. Kervancioglu M, Tokel K, Varan B, Yildirim SV (2011) Frequency, origins and courses of anomalous coronary arteries in 607 Turkish children with tetralogy of Fallot. *Cardiol J*, 18: 546–551.
  10. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L (1979) Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. *Radiology*, 130: 39–47.
  11. Maron BJ (2003) Sudden death in young athletes. *N Engl J Med*, 349: 1064–1075.
  12. Namboodiri N, Harikrishnan S, Tharakan JA (2007) Single coronary artery from right aortic sinus with septal course of left anterior descending artery and left circumflex artery as continuation of right coronary artery: a hitherto unreported coronary anomaly. *J Invasive Cardiol*, 19: E102–E103.
  13. Ongen JA, Goodyer AV (1970) Patterns of distribution of the single coronary artery. *Yale J Biol Med*, 43: 11–21.
  14. Pelliccia A (2001) Congenital coronary artery anomalies in young patients: new perspectives for timely identification. *J Am Coll Cardiol*, 37: 598–600.
  15. Sato Y, Inoue F, Kunimasa T, Matsumoto N, Yoda S, Tani S, Takayama T, Uchiyama T, Tanaka H, Furuhashi S, Takahashi M, Koyama Y, Saito S (2005) Diagnosis of anomalous origin of the right coronary artery using multislice computed tomography: evaluation of possible causes of myocardial ischemia. *Heart Vessels*, 20: 298–300.
  16. Sayar N, Terzi S, Akbulut T, Bilsel T, Ergelen M, Orhan L, Cakmak N, Yesilcimen K (2005) Single coronary artery with subsequent coursing of right coronary artery between aorta and pulmonary artery: fractional flow reserve of the anomalous artery guiding the treatment. *Int Heart J*, 46: 317–322.
  17. Sheth M, Dovnarsky M, Cha SD, Kini P, Maranhao V (1988) Single coronary artery: right coronary artery originating from distal left circumflex. *Cathet Cardiovasc Diagn*, 14: 180–181.
  18. Shirani J, Roberts WC (1993) Solitary coronary ostium in the aorta in the absence of other major congenital cardiovascular anomalies. *J Am Coll Cardiol*, 21: 137–143.
  19. Taylor AJ, Byers JP, Cheitlin MD, Virmani R (1997) Anomalous right or left coronary artery from the contralateral coronary sinus: “high-risk” abnormalities in the initial coronary artery course and heterogeneous clinical outcomes. *Am Heart J*, 133: 428–435.
  20. Taylor AJ, Rogan KM, Virmani R (1992) Sudden cardiac death associated with isolated congenital coronary artery anomalies. *J Am Coll Cardiol*, 20: 640–647.
  21. Tuncer C, Batoryaliev T, Yilmaz R, Gokce M, Eryonucu B, Koroglu S (2006) Origin and distribution anomalies of the left anterior descending artery in 70,850 adult patients: Multi-center data collection. *Cathet Cardiovasc Int*, 68: 574–585.
  22. Venturini E, Magni L (2011) Single coronary artery from the right sinus of Valsalva. *Heart Int*, 6: e5.
  23. Yamanaka O, Hobbs RE (1990) Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn*, 21: 28–40.
  24. Yoda S, Sato Y, Matsumoto N, Tani S, Takayama T, Nishina H, Uchiyama T, Saito S (2005) Incremental value of regional wall motion analysis immediately after exercise for the detection of single-vessel coronary artery disease: study by separate acquisition, dual-isotope ECG-gated single-photon emission computed tomography. *Circ J*, 69: 301–305.
  25. Zhang LJ, Yang GF, Huang W, Zhou CS, Chen P, Lu GM (2010) Incidence of anomalous origin of coronary artery in 1879 Chinese adults on dual-source CT angiography. *Neth Heart J*, 18: 466–470.