

The prevalence of coronary artery anomalies with coronary computed tomography

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Received: 15.02.2016 • Accepted/Published Online: 02.06.2016 • Final Version: 27.02.2017

Background/aim: In this study, the aim is to investigate the prevalence of various coronary artery anomalies (CAAs), evaluated by 256-detector row coronary computed tomography (CCT), within the Turkish population.

Materials and methods: We included 2973 patients who had received CCT. The high take-off right coronary artery (RCA) and left coronary artery (LCA), the RCA arising from the left sinus (LS), the LCA arising from the right sinus (RS), the circumflex artery (CX) arising from the RS, the single coronary ostium originating from the RS, the CX arising from the RCA, and the RCA arising from the pulmonary artery were evaluated.

Results: Seventy-nine patients (2.65%) were diagnosed with CAAs. The frequency of high take-off of the RCA, LCA, and both was 0.1%, 0.67%, and 0.06%, respectively. Frequency of separate ostium of the left anterior descending artery and CX was 0.67%. The frequency of RCA originating from the LS, CX originating from the RS, LCA originating from the RS, LCA and RCA originating from the pulmonary artery, and CX originating from the RCA was 0.50%, 0.10%, 0.23%, 0.03%, and 0.20%, respectively.

Conclusion: In this study, CCT was demonstrated to be a high-quality imaging modality for the detection of CAAs.

Key words: Coronary artery anomalies, coronary computed tomography, coronary angiography

1. Introduction

Coronary artery anomaly (CAA) is a rare congenital abnormality of the coronary arteries. Although CAA is congenital, it generally tends to be detected by coronary angiography or in autopsies. Various studies have reported it to have a genetic basis, and retrospective research conducted on CAA has indicated its incidence to be between 0.6% and 1.3%. The results from the large-scale case series of Yamanaka and Hobbs were in agreement with these findings, reporting the incidence of CAA to be 1.3% (1–3).

Although CAA is routinely incidentally diagnosed during an invasive coronary angiography (ICA), early diagnosis is important in variants of the disorder that are life-threatening. Life-threatening risks are highest in cases where the coronary artery has an interarterial course and supplies the left coronary artery (LCA) area. Life-threatening symptoms such as arrhythmia, syncope, myocardial infarction, or sudden cardiac death develop in

20% of CAA patients. CAA is the second most common cause of sudden cardiac death among young athletes (3–6).

Coronary computed tomography (CCT) has been increasingly used by medical practitioners due to its good temporal and spatial imaging and noninvasive nature (7). However, although CCT is being administered more frequently, conflicting results have emerged about the incidence of CAAs in terms of CCT. The aim of this study was to investigate the prevalence of CAAs in patients screened by CCT.

2. Materials and methods

2.1. Study design and population

A total of 3000 patients who had undergone a CCT screening for investigation of coronary artery disease or suspicion of CAA in the Department of Cardiology and Radiology at Atatürk University between November 2013 and May 2016 were enrolled in this retrospective study.

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2.2. Inclusion criteria

Patients with suspected coronary artery disease (atypical chest pain, angina-equivalent symptoms, or multiple cardiovascular risk factors) and/or suspected coronary anomaly in the angiography were included in the study. All coronary and cardiac structures of the patients were examined in detail.

Patients whose image quality on the CCT was poor due to high heart rate ($n = 27$) were excluded, resulting in a total of 2973 patients being included in the study. The study was carried out with the approval of the local ethics committee.

2.3. Coronary computed tomography

CCT examinations were performed with a multidetector computed tomography (MDCT) 256-slice dual energy CT device (Somatom Definition Flash, Siemens Healthcare, Forchheim, Germany). A single dose of oral β -blocker (metoprolol) was administered 1 h prior to performing the operation on those patients whose heart rate was greater than or equal to 70 beats/min if there were no contraindications. Iopromide (70 mL, Ultravist 370, 370 mg/mL, Bayer Schering Pharma, Berlin, Germany) followed by 60 mL of serum physiologic were administered through the antecubital vein at a rate of 5 mL/s. The operation began by inserting the bolus tracking medium into the left ventricle. Electrocardiography (ECG) monitoring of the patients was performed and recording was conducted using either the prospective ECG-gated or the retrospective low pitch ECG-gated screening model, depending on the heart rate. The following recording protocols were used: for extremely obese patients with 100 kV or 120 kV tube voltage (body mass index >30); gantry rotation time, 270 ms; slicers, $2 \times 128 \times 0.625$ mm; pitch, 0.23; and tube current, 320 mAs. CCT data were best formed using the R-R range for systolic and diastolic phases, with movement of 0.75 mm and 0.4 mm

slice thickness. All images were transferred to an offline workstation (Syngo Multimodality Workplace Siemens, Siemens, Erlangen, Germany) and then analyzed for consensus by 2 observers, one with 15 years and the other with 4 years of CCT experience, who were blinded to all patient data.

2.4. Definition of coronary anomalies

- High take-off coronary artery: The right coronary artery (RCA) or LCA arising from the ascending aorta above the sinotubular junction.
- RCA originated from left sinus of Valsalva: RCA originated from the left system with a separate ostium.
- Circumflex artery (CX) originated from right sinus of Valsalva: CX artery originated from right sinus of Valsalva with a separate ostium.
- LCA originated from right sinus of Valsalva: LCA originated from right sinus of Valsalva with a separate ostium.
- Separate ostia for left anterior descending artery (LAD) and CX: Left sinus of Valsalva separated by two different ostia with an absence of the LCA.
- CX originated from distal part of RCA: CX originated from the distal part of the RCA.
- Coronary artery originated from pulmonary artery: RCA or LCA originated from the pulmonary artery.

3. Results

A total of 2973 patients were screened retrospectively. The baseline characteristics of the patients are summarized in the Table. CAA was diagnosed in 79 patients (2.65%), and the mean age of the patients was 56 ± 12.1 years. High take-off RCA was seen in 3 patients (0.1%), high take-off LCA in 20 patients (0.67%), and high take-off RCA and LCA in 2 patients (0.06%).

The origination of the LAD and the CX from the left sinus of Valsalva with separate ostium was observed in

Table. Baseline characteristics of patients.

Anomaly type	Number of patients (n)	Rate (%)
High take-off coronary artery	23	0.77
Separate ostia for LAD and CX inside the left SoV	20	0.67
RCA from left SoV	15	0.5
CX originating from right SoV	3	0.1
LCA originating from right Sov	7	0.23
CX originating from RCA distal	6	0.2
Coronary artery originating from pulmonary artery	2	0.06

CX: Circumflex, LCA: left coronary artery, LAD: left anterior descending, RCA: right coronary artery, SoV: sinus of Valsalva.

20 patients (0.67%) (Figure 1). In 15 patients (0.5%), the RCA originated from the left sinus of Valsalva (Figure 2), while in 3 patients (0.1%), the CX originated from the right sinus of Valsalva. A single CAA was observed in 2 patients (0.06%) (Figure 3). In 7 patients (0.23%), the LCA originated from the right sinus of Valsalva, in 1 patient (0.03%) the LCA originated from the pulmonary artery, and in 1 patient (0.03%) the RCA originated from the pulmonary artery. Finally, in 6 patients (0.2%), the CX originated from the distal RCA.

4. Discussion

In this study, CCT was demonstrated to be a reliable method for diagnosing CAAs and showing their anatomic localization and for revealing the course of the coronary arteries as well as their relation with the other cardiac structures and anomalies. Moreover, compared to ICA, CCT involves fewer procedure-related complications and is a more cost-effective method as it does not require hospitalization.

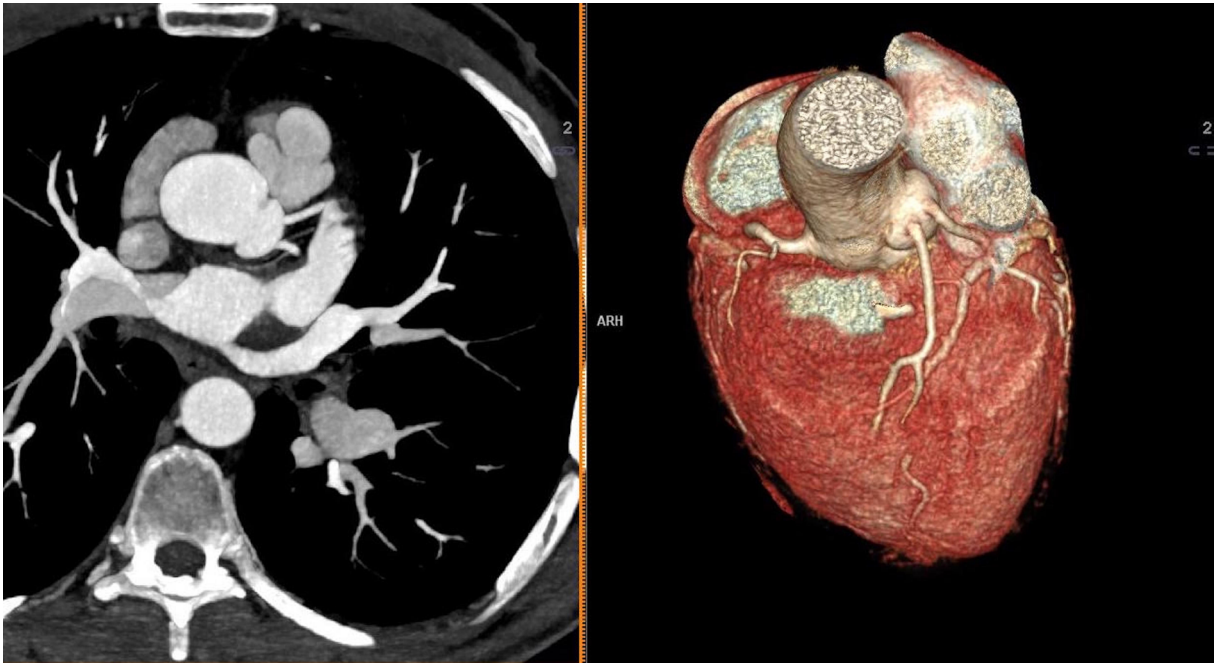


Figure 1. Separate ostia for left anterior descending artery (LAD) and circumflex artery CX.

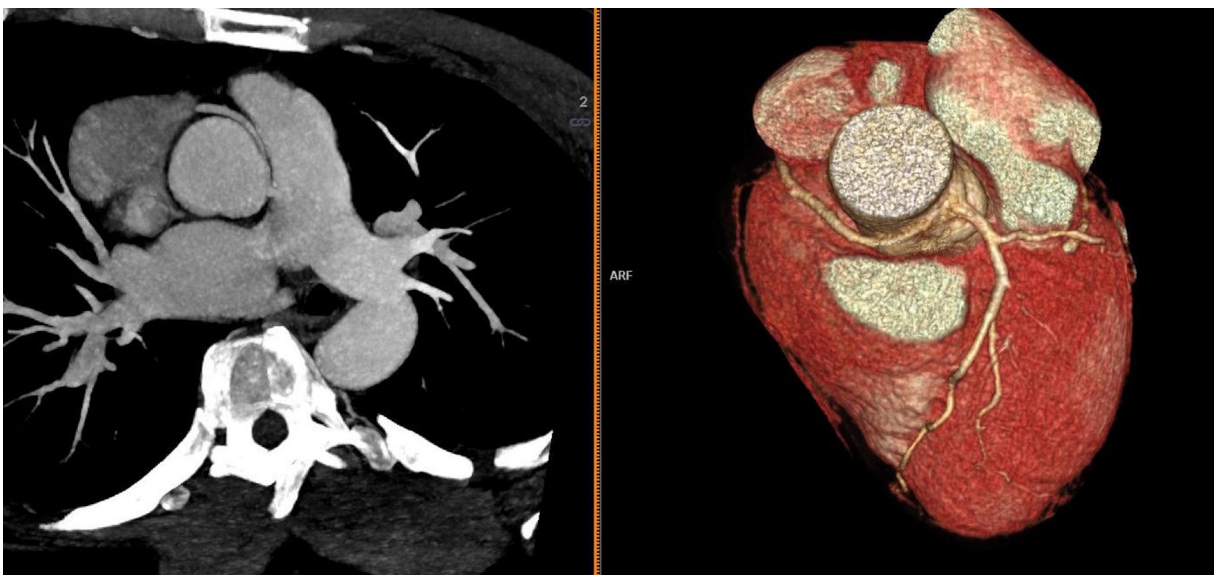


Figure 2. Right coronary artery (RCA) originating from left sinus of Valsalva without interarterial course.

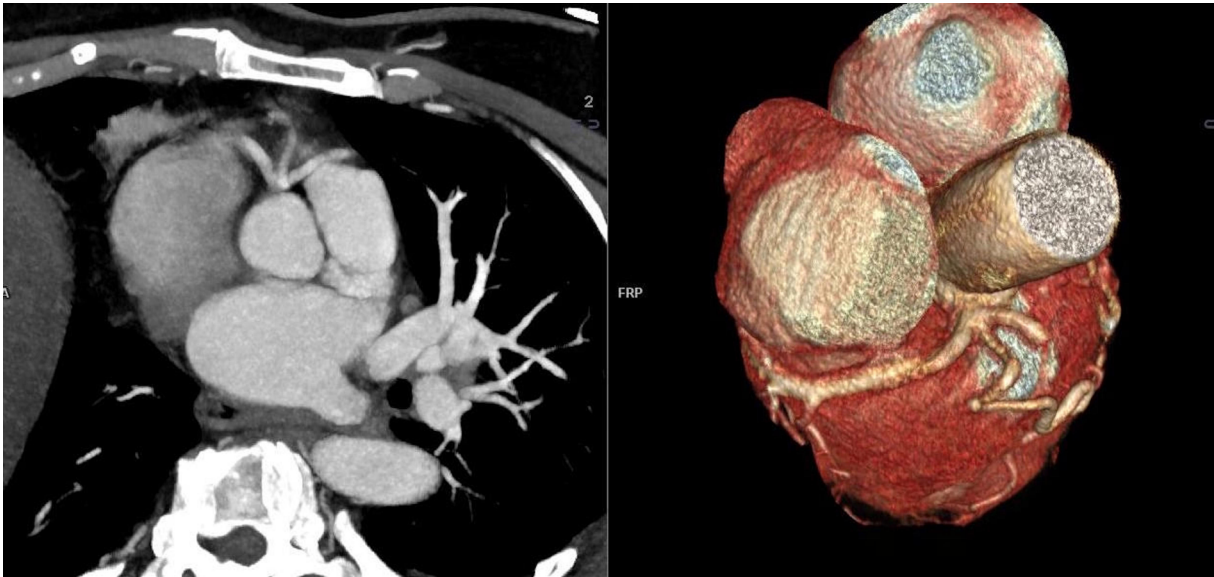


Figure 3. Single coronary artery originating from left sinus of Valsalva.

CAA is a rarely encountered congenital disorder that affects the origination and course of the coronary arteries. It is often diagnosed incidentally following an ICA and seldom causes clinical problems, although in some cases it can result in hemodynamic issues, myocardial perfusion complications, or accelerated atherosclerosis. CAA has an incidence rate of 5.1% in an unselected patient population (7). In one study, such coronary anomalies were found to account for approximately 19% of sudden cardiac deaths in young athletes (8).

High take-off coronary artery is the most frequently observed CAA, with high take-off RCA in particular being the most common. This anomaly is characterized by the coronary artery arising above the sinotubular junction. The less frequently seen types of CAAs include those of the coronary arteries that arise from the aortic arc or the brachiocephalic artery, the internal mammary artery, the bronchial artery, the subclavian artery, and the descending aorta (2,9). In this study, 0.7% of the patients were detected to have either right or left high take-off coronary artery (3 patients RCA, 20 patients LCA). This anomaly is not usually associated with significant hemodynamic problems but can pose difficulties when attempting to cannulate the vessels during ICA. Additionally, when performing aortic surgery, the cross clamping of the aorta below a high take-off artery might result in inadequate induction of cardioplegia (8,10). The projection of the LAD and CX from the left sinus of Valsalva, separately without the LCA, is the second most commonly observed CAA (0.5%–0.8%) and was observed in 20 patients (0.67%) in this study's patient group. In this anomaly, the LCA is not observed and the CX and LAD arise from two separate ostia in the sinus of Valsalva. These cases are not associated with

any clinical events, except those related to cannulation problems (7).

The origination of the coronary artery from the other ostium is defined by the RCA arising from the left sinus of Valsalva, the LCA arising from the right sinus of Valsalva, and the CX artery arising from the right sinus of Valsalva. In this anomaly, which is diagnosed as a result of the coronary artery being unable to be cannulated during an ICA, the coronary arteries have a tendency to run an interarterial course. Therefore, when the diagnosis is made, this feature in particular needs to be further examined. With a prevalence ranging from 0.2% to 1%, the anomalous origination of a coronary artery from the opposite sinus of Valsalva is one of the most common causes of sudden cardiac death in young people and is usually observed to occur after strenuous exercise (3,7,8). In this study, the RCA originated from the left sinus of Valsalva in 15 patients (0.5%), the CX originated from the right sinus of Valsalva in 3 patients (0.1%), and the LCA originated from the right sinus of Valsalva in 7 patients (0.23%). CCT was performed on these patients to determine, via the three-dimensional structure produced by the CCT, the location from which the anomaly originates and to evaluate the proximal middle and distal portion of the coronary arteries and the relation between the coronary arteries and other cardiac structures. Once these data are ascertained, the risk of sudden death can be determined (9). In this study's series, 6 patients were found to have a malignant interarterial course of the anomalous RCA and LAD (0.20%). The RCA originating from the left sinus of Valsalva ordinarily shows intraarterial circulation and is related to sudden death. This malignant RCA anomaly was detected in 5 patients in the study and is consistent

with the literature (3,8,9,11). Another type of CAA, the CX artery originating from the RCA distal portion, was observed in 6 patients (0.20%). This anomaly is defined by the CX artery arising from the distal part of the RCA yet still anatomically supplying blood to the area that is supplied by the CX artery in normal circumstances, and by the absence of the CX artery in the ICA. In this anomaly, the CX shows normal distribution, there are no malignant variations, clinical symptoms are absent, and the condition is accepted as benign (8,12). A single coronary artery defines coronary arteries arising from a single aortic ostium. Only rarely observed, this anomaly causes cannulation problems during ICA and constitutes an increased risk for sudden death due to it running an interarterial course. In this study, it was observed in 2 patients (0.06%), with it originating from the right sinus of Valsalva in 1 patient (0.03%) and from the left sinus of Valsalva in the other patient (0.03%). In both of these patients, it was determined to be a malignant variation.

The coronary artery originating from the pulmonary artery can be defined as one of the most serious congenital coronary anomalies. In this study, the LCA originated from the pulmonary artery in 1 patient (0.03%), while the RCA originated from the pulmonary artery in 1 patient (0.03%). Although not observed in this study, CX or any coronary arteries originating from the pulmonary artery have only rarely been reported. This anomaly can either be asymptomatic or may occur together with other cardiac anomalies (tetralogy of Fallot, atrial septal defect, ventricular septal defect, and aortic coarctation). One of the specific findings of this disease is the development of prevalent tortuous dilate collaterals, and in connection with such collaterals, the occurrence of coronary steal from the high-pressure coronary arterial system to the low-pressure pulmonary artery. In cases where the RCA arises from the pulmonary artery, no symptoms of stealing appear due to the low pressure naturally occurring in the right ventricle and clinical treatment is tolerated better; however, in cases where the LCA arises from the pulmonary artery, ischemia develops due to the high pressure of the left ventricle. The ischemia triggers collateral development as well as the development of tortuous collaterals, and coronary steal syndrome develops from the high-blood-pressure coronary system to the low-blood-pressure pulmonary arterial system. Surgical treatment is usually recommended for these patients (2,3,10,11).

ICA traditionally served as the primary choice for detection of CAAs. However, because ICA was unsatisfactory in terms of imaging the course of the coronary artery system and its relationship with other anatomical structures, of showing the other cardiac structures, and of having the capacity to reveal anomalies simultaneously, other alternative imaging techniques have

begun to be used. As a result of the advanced developments made in CCT in recent years and CCT's better image quality, ability to produce simultaneous imaging with other cardiac structures, and low radiation exposure, CCT imaging has been extensively used in this field as an alternative method (5,9,12).

In CCT studies, the CAA incidence was found to be higher than in ICA studies (13,14). De Jonge et al. reported a CAA prevalence of 7%, which included coronary fistulas, in their patient population (15). In one study, the prevalence of CAAs of origin and further course was reported to be 7.9% in mainly symptomatic patients (13). Finally, CAA incidence was found to be 0.8% in the series study recently performed by Srinivarsan et al. (14). According to the risk scoring system, these figures can be explained by the fact that the high CAA incidence in the young group was the result of ICA being used to evaluate the middle-aged and elderly group, while MDCT was used to evaluate the young group. Nonetheless, CAA incidence tends to be significantly higher with respect to ICA. Despite these conflicting results shown in the studies, the figures produced often do not reflect the real incidence. This situation could be related to the fewer occurrences of congenital anomalies with respect to CCT, and to the inability of the angiographer, who must rely on the ICA, which offers only two-dimensional imaging, to perform correct categorization. On the other hand, the three-dimensional structure produced by CCT is able to provide more detailed information to overcome this problem and thus can explain the high incidence in CCT studies (7,9,11,13–17). Finally, compared with ICA, CCT requires less skill and experience, has lower radiation exposure, and poses no risks for bleeding in the puncture point, artery-venous fistula formation, coronary artery dissection, stroke, or death. Moreover, it does not require hospitalization, as it is an outpatient procedure (7).

Another advantage of CCT is that it allows for identification of malignant CAAs, which are related to sudden cardiac death. ICA, on the other hand, often fails to identify these. CCT has therefore become an important method, with its ability to generate a three-dimensional structure and its low radiation exposure, as recently attested to by the American College of Cardiology, which recommended the use of CCT for evaluation and cases of CAA suspicion (2,3,17,18).

It should be noted that our study was retrospective and therefore did not make a direct comparison of two different modalities. The study consisted of patients suspected of coronary artery disease or CAA who were referred by cardiologists, and because young patients in particular commonly undergo evaluations using CCT rather than ICA, the higher CAA incidence with respect to the invasive method could be related to selection bias.

In conclusion, this study provides important evidence of the superiority of CCT over ICA, in terms of defining CAAs and classifying clinical risks, due to the former's ability to produce high-quality three-dimensional imaging, to show the relationship between anatomic structures, to

cause fewer complications, and to require fewer cases of hospitalization. Knowing the CCT properties of CAAs and their clinical importance will help to facilitate a correct diagnosis and a suitable treatment plan.

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