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Rare diseases of systemic circulation

The prevalence of coronary artery anomalies on CT scan – experience from a tertiary care center in Pakistan (RCD code: I-1C.1)

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Abstract

Objectives: This study is an effort to bridge the gap regarding the data of prevalence of coronary anomalies in South Asian countries by utilizing coronary computed tomography angiography (CCTA). Methods: This is a cross sectional, descriptive study done at Aga Khan University Hospital Karachi, Pakistan. All adult patients who underwent CCTA from 1-1-2005 to 1-11-2016 were included in the study. Results: Among 897 patients undergoing CCTA, 36 (4.01%) patients were identified as having coronary artery anomalies (CAA). The mean age of patients was 48.3±11.9 years. Male to female ratio was 3:1. Among patients with CAA, the most common indication for CCTA was screening for coronary artery disease (58.3%). 33 patients (3.67%) showed coronary origin and course anomalies, whereas coronary artery fistulas were identified in 3 patients (0.3%). The most common anomaly identified was the anomalous origin of coronary artery from the opposite coronary sinus with anomalous course (1.4%) with anomalous origin of right coronary artery from the left coronary sinus with an inter-arterial course seen in 9 patients. Single coronary artery was seen in three cases (0.3%), in all these cases the single coronary artery originated from the right coronary cusp. In one patient (0.1%) dual left anterior descending (LAD) system was identified. In two cases (0.2%) fistulous connection of LAD with pulmonary artery was seen, while fistula from LAD to coronary sinus was identified in one patient. Conclusions: This study highlights the fact that the prevalence of coronary anomalies on CCTA in this region is similar to the data reported from the rest of the world. JRCD 2017; 3 (4): 116–121

Key words: rare disease, coronary anomalies, prevalence, coronary CT angiography, tertiary care center

Introduction

The prevalence of anomalous coronary arteries in general population is reported to be around 0.3–37% [1–10]. Coronary artery anomalies (CAA) can occur as an abnormal origin, atypical course or unusual termination. These anomalies are mostly detected as incidental findings on conventional coronary angiography (CCA) and are in most cases benign. Seldom patients present with cardiac symptoms because of anomalous coronary artery origin or termination. Yet, potentially lethal variants of coronary anomalies have also been recognized.

The introduction of coronary computed tomography angiography (CCTA) has provided a non-invasive and convenient method of visualization of coronary anatomy. With the increased usage of CCTA and the advancement in the quality of image acquisition the detection of anomalies of origin, course and termination of coronary arteries has greatly improved. The CCTA has been reported superior to conventional angiography for defining anomalous coronary artery origin [11]. CAA can be classified on the basis of anomalies of origin, course of the vessel and termination using a modified theme presented by Greenberg et al. [12].

Data regarding the prevalence of CAA detected on CCTA has been reported, but there is lack of such data from Pakistan. The probable reasons includes lack of facilities for performing and reporting CCTA.

In order to bridge this gap, we performed a retrospective analysis of all CCTA that have been performed in a CCTA equipped tertiary care center in Pakistan and we present our results regarding the prevalence of CAA including anomalies of origin, course and termination.

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Materials and methods

This was a cross sectional study conducted at the Aga Khan University Hospital Karachi from 1st September 2016 to 30th April 2017. As this was a retrospective analysis no sample size was required. The study included all consecutive, adult patients, aged 18–80 years undergoing CCTA from 1-1-2005 to 1-11-2016 for the evaluation of symptoms of chest pain, syncope, dyspnea or palpitations, preoperative evaluation for surgery, for screening of coronary artery disease and for determination of the patency of bypass grafts or stents. Patients undergoing computed tomography (CT) for the assessment of complex congenital heart disease were excluded.

The study was commenced after taking ethical approval from the ethical review committee of the hospital. All patients had signed informed consent to be a part of this study. Data of all patients undergoing CCTA fulfilling the inclusion criteria at the Aga Khan University Hospital were reviewed from the hospital medical records system. CT scan reports were reviewed using the patient profile viewer software and the data was filtered to record prevalence of coronary artery anomalies among patients. The demographic data of the patients and the indications for performing CCTA were recorded.

The primary outcome was the prevalence of coronary artery anomalies. All data was analyzed using SPSS version 21.0. Means and standard deviations were calculated for age. Frequency and percentages were calculated for gender, diabetes mellitus, hypertension, dyslipidemia, smoking, prior history of ischemic heart disease and type of coronary artery anomaly.

Data acquisition and reconstruction protocol

All CT examinations were performed on a 64-slice CT scanner (Toshiba). Coronary angiography scan was started by continuously injecting a bolus of 80 ml of non-ionic contrast agent followed by 50 ml saline solution into an antecubital vein via an 18-gauge catheter (injection rate 5 ml/s). Contrast agent application was controlled by a bolus tracking technique. A region of interest was placed into the aortic root, and image acquisition started 5 seconds after the signal attenuation reached the predefined threshold of 100 Hounsfield units (HU). A retrospective gating technique was used to synchronize the data reconstruction with the electrocardiography (ECG) signal. In each patient, 10 CT data were reconstructed in 10% steps of the R-R interval. Images were reconstructed with a slice thickness of 1.0 mm, a reconstruction increment of 1.0 mm, and using a medium soft-tissue convolution kernel (B26f). Depending on the individual anatomy, the reconstructed field-of-view (FOV) was adjusted to encompass the heart exactly (image matrix 256 × 256 pixels).

Table 1. Baseline characteristics of 36 patients with coronary anomalies

Baseline Characteristics	N (%)
No. of patients	36
Age (years)*	48.39±11.9
Male	27(75)
Presenting symptoms	
Atypical chest pain	03(8.3)
Dyspnea	01(2.8)
Palpitations	01(2.8)
Screening of coronary artery disease	21(58.3)
For determination of the patency of bypass grafts or stents	02(5.6)
Rule out congenital heart disease	03(8.3)
To confirm presence of anomalous circulation identified on invasive angiogram	04(11.1)
Positive stress test	01(2.8)
Risk factors	
Hypertension	25(69.4)
Dyslipidemia	11(30.6)
Diabetes	08(22)
Smoking	06(16.7)
History of IHD	5(13.9)

IHD – ischemic heart disease

Image reformation and analysis

After removing the patient, all reconstructed images were transferred to a dedicated workstation equipped with dedicated cardiac post-processing software (Vitrea). Image post-processing was performed using techniques of maximum intensity projection (MIP), multiplanar reformation (MPR), and volume rendering (VR) for the optimal phase data.

Results

Among 897 patients undergoing CCTA, 36 (4%) patients were identified as having CAA. The mean age of these patients was 48.3±11.9 years (range 24–77 years). Male to female ratio was 3:1. Among these 36 patients, the most common indication for CCTA was screening for coronary artery disease (21 patients, 2.3%). The baseline characteristics of the patients are shown in Table 1. Among patients with CAA, 33 persons (91.6%) showed coronary artery origin and course anomalies, whereas coronary artery fistulas were identified in 3 patients (8.3%), Table 2. The most common

Table 2. Prevalence of coronary anomalies

Coronary anomaly	Number of patients	Anomaly incidence among 897 patients (%)
Origin and Course anomalies	33	3.67
Anomalous origin from opposite coronary sinus with anomalous course		
RCA arising from left coronary sinus with interarterial course	09	1.00
LCx arising from the right coronary sinus with retroaortic course	02	0.2
LCx arising from the right coronary artery with retroaortic course	01	0.1
Origin of LCA and RCA from a fistulous connection between the right coronary sinus and left aorto-atrial tunnel	01	0.1
Single coronary artery arising from right coronary sinus with interarterial course of LAD	01	0.1
Single coronary artery arising from right coronary sinus with retroaortic course of LCA	01	0.1
Single coronary artery arising from right coronary sinus with prepulmonic course of LCA	01	0.1
Separate origin of LAD and LCx(absent LM trunk)	04	0.4
Duplication of arteries (type IV dual LAD)	01	0.1
Low takeoff of LCA	03	0.3
Myocardial bridging	09	1.0
Coronary termination anomalies (coronary artery fistula)	03	0.3
LAD to PA	02	0.2
LAD to CS	01	0.1

RCA – right coronary artery, LCx – left circumflex coronary artery, LCA – left coronary artery, LAD – left anterior descending artery, PA – pulmonary artery, CS – coronary sinus

anomaly identified was the anomalous origin of coronary artery from the opposite coronary sinus with anomalous course seen in 13 patients (1.4%). Nine patients (1%) showed anomalous origin of right coronary artery (RCA) from the left coronary sinus (Figure 1) with an inter-arterial course between the aorta and the pulmonary trunk. Two patients (0.22%) had origin of left circumflex coronary artery (LCx) from right coronary sinus and one patient from RCA (0.1%). In all these cases, the LCx had a retro aortic course after origin. In one case of a 24-year-old man, anomalous origin of left coronary artery (LCA) and RCA was noted from a fistulous connection between the right coronary sinus and left aorto-atrial tunnel. Single coronary artery was seen in three cases (0.3%), in all these cases the single coronary artery originated from the right coronary cusp (Figure 2). In one patient (0.1%) dual left anterior descending (LAD) system was identified, one LAD arising from left coronary cusp and coursing in the anterior interventricular groove with the other LAD arising from the RCA, and coursing anterior to the pulmonary artery. In the same case, LCx also originated from RCA, and coursed between the aorta and left atrium. Four patients (0.44%) had no left main coronary artery (LMCA) with a separate origin of LAD and LCx from the left coronary sinus. Three patients (0.33%) had low take off of LCA from the left coronary sinus. Coronary artery fistulas were seen in 3 patients (0.3% of cases). In two cases (0.2%) fistulous connection of LAD with pulmonary artery was seen (Figure 3), while fistula from LAD to coronary sinus was identified in one patient. Myocardial bridging was noted in 9 patients (1%) with mid LAD involvement in all cases. Among the 36 patients identified as having coronary anomalies on CT angiogram, all patients were doing well on telephonic follow-up and no patients had major cardiovascular events, including sudden cardiac death related to the coronary anomalies.

Discussion

The prevalence of anomalous coronary circulation is highly variable 0.3–37% [1–10]. This discrepancy is likely due to the lack of an accepted definition of anomalous coronary circulation and referral bias in the populations studied. The prevalence of CAA in our study is 4%. This prevalence is higher than the reported prevalence of CAA on conventional angiography, yet within the prevalence reported on CCTA studies. A review of studies with higher reported prevalence shows that due to lack of clear definition of

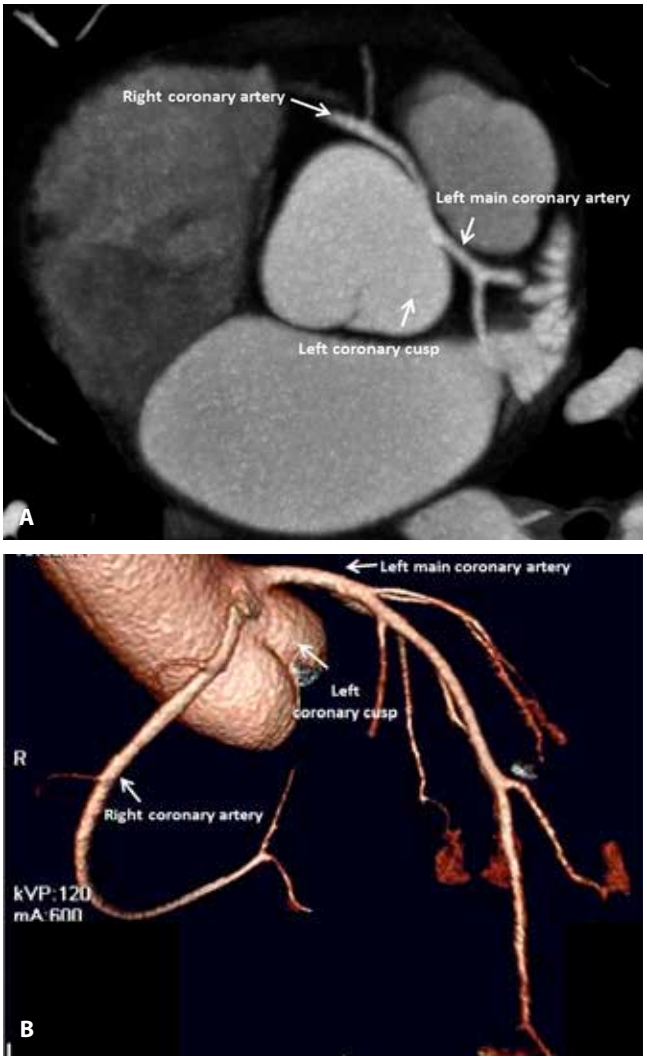


Figure 1. A. Anomalous origin of right coronary artery from left coronary cusp. B. 3-D reconstruction showing anomalous origin of right coronary artery from left coronary cusp

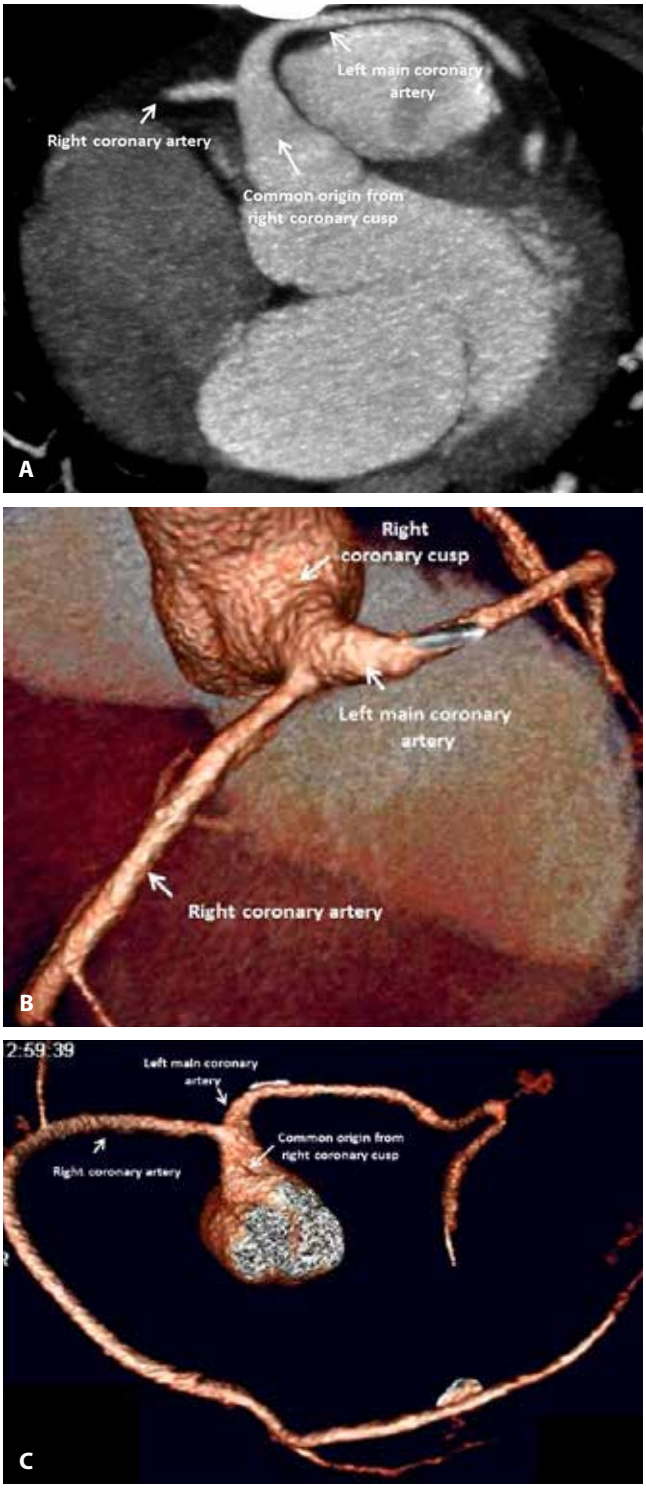
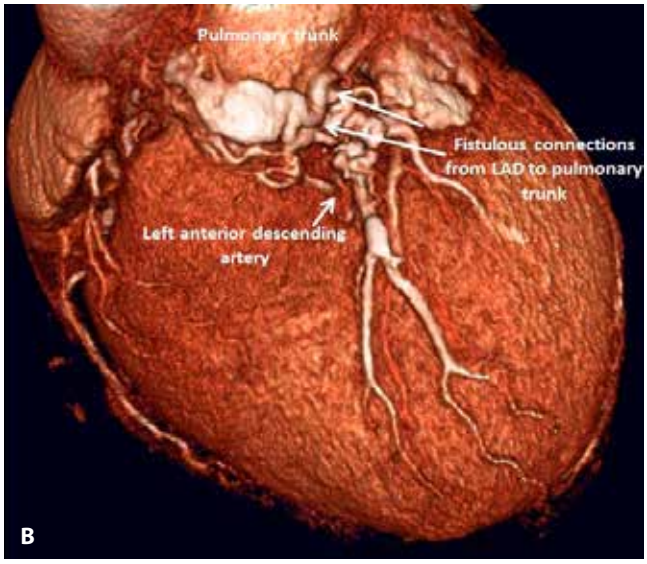


Figure 2. A. Anomalous common wide origin of right coronary artery and left coronary artery from right coronary cusp. B–C. 3-D reconstruction showing anomalous origin of right coronary artery and left coronary artery from right coronary cusp

CAAs these studies included patients with suspected anomalous circulation on conventional angiograms and certain anomalies such as myocardial bridging and fistulas [5,6].

The most common anomaly that was observed in our study was the anomalous origin of coronary artery from the opposite coronary sinus with anomalous course (1.4%). Of these anomalies the most common anomaly was the origin of RCA from the left coronary sinus (1.00%). This is in conjunction with the reported prevalence in general population [7–10]. A coronary artery arising from the opposite or non-coronary sinus can take any of four common courses, depending on the anatomic relationship of the anomalous vessel to the aorta and the pulmonary trunk. These paths include an interarterial course between the aorta and the pulmonary artery, a retroaortic course behind the aorta, a prepulmonic path in front of the pulmonary trunk and a septal (subpulmonic) course beneath the right ventricular outflow tract [13]. The interarterial course has been identified as a potential risk factor for sudden cardiac death [13]. In all our cases of origin of RCA from the left coronary sinus, the RCA had an interarterial course between the aorta and the pulmonary trunk. There was no LMCA with an interarterial course identified in our study, probably because of the high mortality at young age associated with this abnormality. The origin of LCx from the right coronary sinus (RCS) with a retroaortic course is considered a benign anomaly, however, the identification of this



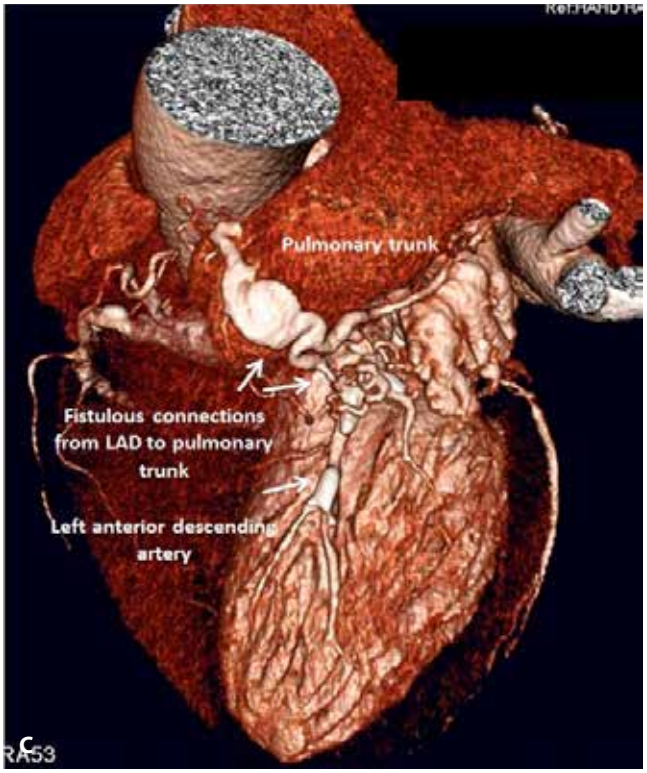


Figure 3. A. Multiple fistulous channels originating from proximal left anterior descending artery, making an aneurysmal sac over the main pulmonary artery and then draining into the main pulmonary artery. B–C. 3-D reconstruction showing the fistulous connections

coronary anomaly is vital for patients planned for prosthetic valve surgery to prevent compression of the LCx during surgery [1,14].

A single coronary artery (SCA) is considered a benign anomaly, however, it is associated with increased risk of sudden death if a branch from SCA crosses between the pulmonary artery and the aorta [15]. The prevalence of single coronary artery is reported to be 0.014–0.066% with the SCA arising more commonly from the right coronary sinus [16]. In our study, 3 cases of single coronary artery were identified with a prevalence of 0.33%. The prevalence of SCA in our study was higher when compared with prevalence reported in other studies.

Another benign abnormality noted in our study was the separate origin of LAD and LCx from the left coronary cusp. The prevalence of separate origin of LAD and LCx is reported to be 0.4% [17]. This abnormality leads to difficulties in vessels cannulation. Identification of this abnormality is essential for patients undergoing cardiopulmonary bypass surgery to selectively graft these separate vessels. The prevalence of this abnormality in our study was 0.44%.

Anomalous terminations of coronary arteries include coronary artery fistulas. This condition is characterized by a communication between the coronary artery and either a cardiac chamber, systemic vein, or the pulmonary artery. Coronary artery fistulas are asymptomatic in adults, and surgical correction is indicated when there is high fistula flow, with multiple communications and significant aneurysm formations [18].

Clinical consequences of coronary artery fistulas depend on their drainage sites, the most common sites of drainage of fistulas is the right ventricle. The prevalence of this anomaly is reported to be 0.05% to 0.25% on conventional angiograms. However, the reported prevalence on CCTA studies is 0.35–0.9% [8,19]. In our study coronary artery fistulas were seen in 3 patients (8.3% of cases) with a prevalence of 0.33%. In all these patients the fistulas was fed by the LAD, in two cases the fistula had a terminal connection with the pulmonary artery, whereas in one patient the fistula terminated in the coronary sinus.

Dual LAD is a benign abnormality with a prevalence of 1% [20] on CCA and 4% [21] on CCTA. In our study one case of dual LAD was identified (prevalence of 0.11%).

The 3D image reconstruction by CCTA has led to better detection of fistula patterns and delineating complex fistula anatomy. The finer image acquisition by high resolution CCTA has improved coronary artery fistula detection on CCTA with a reported prev-

alence of 0.35% in previous studies. The prevalence in our study was similar to previously reported prevalence in CCTA studies and higher as compared to CCA studies.

This is the first study on CCTA and CAA to be reported from Pakistan and our findings are comparable with previous reported studies [5,7–10,17,19,20]. The main limitation of this study is its retrospective design. Another limitation of the study is that the study does not shed light on the follow-up of cases.

Conclusions

CAA are rare but their identification is important as they can result in possible fatal outcomes. With the advancement in the field of CCTA imaging and the introduction of high resolution multi-row detector CT devices that provide finer image quality with a high spatial resolution, the detection of CAA has improved considerably. This enables physicians to correctly identify CAA and guide interventional cardiologists and cardiac surgeons to prevent procedural complications. This study brings to focus the fact that the prevalence of CAA on CCTA is similar to the reported data from the rest of the world. Further work is required in this field to document the outcomes of patients identified to have CAA on CCTA and the clinical significance of these anomalies in asymptomatic patients.

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