

PREVALENCE OF CONGENITAL CORONARY ARTERY ANOMALIES IN ADULTS PATIENTS UNDERGOING CONVENTIONAL CORONARY ANGIOGRAM-A RETROSPECTIVE STUDY IN A TERTIARY CARE HOSPITAL IN TAMIL NADU

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ABSTRACT

Background & Objectives: Coronary artery anomalies (CAAs) are a diverse group of congenital disorders whose manifestations and pathophysiological mechanisms are highly variable. Among patients undergoing coronary angiography, the incidence depends on the population studied and the criteria used to define an anomaly. This study is conducted to study the prevalence and pattern of coronary anomalies in our geographic region.

Methods: We retrospectively studied 3000 coronary angiographies (CAG) done in the institution from JANUARY 2017 to JUNE 2020 for the detection of incidence and pattern of coronary anomalies. Patients with ischemic heart disease and valvular heart disease who underwent CAG were included in the study.

Results: Total prevalence of CAA is 2.4% as per Angelini's classification and 1.1% as per modified Angelini's classification. The most common anomaly was absent left main artery with separate origin of the left anterior descending (LAD) artery and left circumflex artery (LCx) (n=18, 0.6%). Anomalous origin of LCx from right sinus/ RCA was seen in 4 patients (0.13%). Anomalous origin of the right coronary artery (RCA) from left circulation was seen in 4 patients (0.13%) and directly from the aorta in one patient. One patient had an aberrant common trunk from the right sinus of Valsalva, with the common trunk bifurcating into the right and left system shortly after its origin. Atretic coronary artery and origin of the coronary artery from the pulmonary artery were not seen in our study.

Conclusion: Although CAAs are rare causes of sudden death, the anomaly of a single coronary artery, an LMCA originating from the pulmonary artery, and congenital hypoplastic, stenotic, or atretic LMCA may lead to myocardial infarction and sudden death. Though in the present study we didn't find such malignant anatomy, the total prevalence of coronary anomalies was similar to that in other studies.

KEYWORDS: Coronary anomalies, coronary anatomy, variation, the incidence

INTRODUCTION

Coronary artery anomalies (CAAs) are a diverse group of congenital disorders whose manifestations and pathophysiological mechanisms are highly variable. With an increasing number of coronary angiography (CAG) procedures, coronary invasive procedures, and cardiac bypass surgeries performed every day, knowledge about the variations in the anatomical pattern of coronary arteries is pivotal to avoid complications. [1] The widespread application of coronary angiography (CAG) has resulted in congenital coronary artery anomalies being identified more frequently and an improved understanding of the clinical significance of such anomalies. [2] Angiographic recognition of coronary anomalies is of great importance. A surgeon must know the presence of coronary anomalies of origin and distribution, to perform optimal cardioplegia and not to cut an anomalous artery. It is similarly crucial for an angiography operator to be aware of coronary anomalies in order not to miss an anomalous vessel. Failure to identify these anomalies can lead to inadequate and prolonged procedures, which can result in catastrophic complications. [3] Coronary arteries are broadly categorized into three groups based on their anatomical features: normal coronary anatomy, anatomic variations of the coronary artery, and

coronary artery anomalies (CAAs). Usually, there are two coronary ostia giving rise to two main coronary arteries that supply oxygenated blood to the myocardium – the left main coronary artery (LMCA) originating from the left sinus of Valsalva (SV) and the right coronary artery (RCA) originating from the right SV. There is no artery originating from the posterior SV which is otherwise called the Noncoronary cusp (NCC). After originating, the LMCA bifurcates into the left anterior descending (LAD) artery which runs in the anterior interventricular sulcus, providing the penetrating septal branches, and the Left circumflex artery (LCX) which runs along the left atrioventricular sulcus and gives rise to at least one obtuse marginal (OM) branch.[4] The RCA lies in the right atrioventricular sulcus and gives rise to the acute marginal branch. At times there maybe an additional artery called the intermediate artery (IMA) or Ramus intermedius forming a trifurcation of LMCA. IMA is located anterior to the first OM artery and posterior to the first diagonal artery supplying the left ventricular free wall. The artery that supplies the posterior descending artery determines coronary dominance. Approximately 70%–80% of the general population is right-dominant (i.e. supplied by the RCA), while 5%–10% is left-dominant (i.e. supplied by the CXA) and 10%–20% is co-dominant (i.e. supplied by both the RCA and CXA). A more accurate definition of dominance refers to the arterial supply to the atrioventricular nodal artery, which is generally supplied by the RCA.[5] However, in some cases where the LMCA is absent, three ostia can be detected. In individuals with such a condition, the LAD artery and CXA originate from different ostia. The absence of the LMCA is a common anomaly that can be detected in 0.4%–8% of the population. [6] Angelini identified a set of criteria that allowed the systematic assessment of coronary arteries and hence the description of normal coronary anatomy. Angelini introduced the concept of normal variant versus anomaly based on a statistical definition of what constitutes the normal range (coronary pattern observed in 99% of an unselected population) and CAAs (coronary patterns observed in less than 1% of the study population). This systematic anatomic approach is probably the most valid classification method for CAA. (7) Generally evaluated using CAG, Coronary anomalies are observed in less than 1% of the general population. [8] A myocardial bridge (MB) is a condition in which a coronary artery tunnels through the myocardium rather than resting on top of it. A CAF is an abnormal connection between a coronary artery and another structure, most commonly a venous structure or a chamber on the right side of the heart. Abnormal dilatation of a part of the coronary artery is called coronary artery ectasia or coronary aneurysm. Coronary artery ectasia is the dilatation of a coronary artery segment to a diameter 1.5 to two times that of the adjacent segment, whereas coronary aneurysm is the dilatation of a coronary artery segment to a diameter more than two times that of the adjacent segment [9]. The incidence of coronary anomalies in the adult population varies on angiographic and postmortem studies. Although generally benign, some coronary artery anomalies are associated with more serious clinical outcomes, such as congestive heart failure, arrhythmia, myocardial infarction, syncope, and sudden death. The coronary dominance pattern, presence of IMA (Intermediate or Ramus artery), and incidence of CAAs may vary depending on the population studied. In the present study, we analyze coronary angiograms to determine the frequency and types of coronary artery origination and course anomalies in our center. [10]

METHODS:

It's a retrospective study of Totally 3000 patients both Male & Female undergoing coronary angiography in SRM institute of medical sciences & research in the Department of Cardiology during the period of 1/1/2017 to 30/6/2020 were included in the study. The coronary angiograms of the involved patients will be analyzed retrospectively and searched for any congenital coronary anomalies. The clinical presentation of the patients detected to have coronary anomalies will be reviewed. The Coronary dominance pattern, the presence of IMA and CAA were recorded. CAAs were grouped according to the systematic anatomic approach developed by Angelini and Khatami, which is recognized as the most valid classification for CAA. (10) A simplified modification of Angelini and Khatami's classification which is currently used in clinical practice divides CAAs into two groups: Group A, anomalies of origin and distribution; and Group B, intracoronary communications and coronary artery fistulae. Both classification systems were used to evaluate the CAAs in the present study. **INCLUSION CRITERIA:** Patients undergoing coronary angiography in the above set period. **EXCLUSION CRITERIA:** Patients with complex congenital heart diseases, high or low "take-off" of coronary arteries, and separate origin of the CONUS artery from the right coronary sinus (RCS).

STATISTICAL ANALYSIS

Statistical analysis was performed using SPSS, version 15.0 (SPSS Inc., Chicago, IL, USA). Results were analyzed with the Student's *t*-test for quantitative data that were normally distributed and with the Mann–Whitney *U*-test for quantitative data that were not normally distributed. Categorical data were analyzed with the chi-square or Fisher's exact test (when Levene's test was significant). *P*-values < 0.05 were considered statistically significant.

RESULTS: During the aforementioned study period, a total of 3000 subjects were recruited into the study, and all the subjects were included in the final analysis. The mean age of the patients who had undergone diagnostic CAG was 55.8 ± 10.4 years. Most of the patients were men constituting around 71.6% (2150) of the study population. The most frequent CAG indications were chest pain, a positive stress test, and wall motion abnormality on echocardiography. There was no

significant difference in the coronary dominance pattern and there was no significant difference between the two genders. IMA was detected in 330 (11.0%) patients without any statistically significant difference between males and females.

TABLE:1 CORONARY DOMINANCE PATTERN

Right Dominant	82.6%
Co-Dominant	11.5%
Left dominant	5.9%

TABLE :1 According to Angelini and Khatami's classification (table 2), 73 patients out of 3000 had congenital coronary anomalies making the overall incidence to be 2.4%. Based on the modified classification, 28 (0.9%) patients had anomalies of origin and distribution (i.e. Group A), while 6 (0.2%) had intracoronary communications and coronary artery fistulae (i.e. Group B). As the modified classification does not accept MB and coronary artery aneurysms as coronary anomalies, the incidence of CAAs was found to be 1.1%. We found the absent LMCA anomaly in 18(0.6%) patients, making it the most common anomaly observed in the present study. Absent LMCA leading to separate origins of LAD and LCX was seen in 0.6% of patients (18 patients). One patient had An aberrant common trunk from the right sinus of Valsalva, with the common trunk bifurcating into the right and left system shortly after its origin. The RCA originated from the left coronary circulation (i.e the left SV or left coronary arteries) in four patients (0.13%) and directly from Aorta in one patient. The LCX originated from the right coronary circulation (i.e the right SV or right coronary arteries) in four patients(0.13%). CAF was detected in 6 (0.2%) patients – all originated from the RCA and drained into RV. Myocardial bridging (MB) was detected in 30 (1.0%) patients and at a higher frequency among the male patients than the female patients (0.8% vs. 0.2%, p = 0.02). Coronary ectasia was observed in 162 (5.4%) patients; a higher frequency was again observed among the male patients than among the female patients. Aneurysms were detected in 13 (0.4%) patients and slow coronary flow was diagnosed in 33 (1.1%) patients. Anomalous origin of the coronary arteries from the pulmonary artery or atretic coronary arteries was not observed in the present study. The most common clinical presentations of the patients with CAA were: atypical chest pain (40%); typical angina (33%); acute coronary syndrome (27%). None of the patients had a history of syncope.

TABLE:2 ANGELINI AND KHATAMI'S CLASSIFICATION

Parameter	Numberof Patients	Angiographic incidence
Anomalous pulmonary origins of the coronary arteries	-	-
Anomalous aortic origins of the coronary arteries	28	0.9%
Myocardial bridge	30	1.0%
Coronary arteriovenous fistulae	6	0.2%
Coronary artery aneurysms	9	0.3%
Total incidence(as per original classification)		2.4%

MODIFIED CLASSIFICATION

Group A (anomalies of origin and distribution)	28	0.9%
Group B (intercoronary communications and coronary artery fistulae)	6	0.2%
Total incidence(as per modified classification)		1.1%

TABLE:2 The mean of several catheters used during CAG was 2.54 ± 0.76 (range 2–4) as shown in Table 3. The mean duration of fluoroscopy was 10.2 ± 3.8 (range 3–24) minutes.

TABLE :3 NUMBER OF CATHETERS USED DURING CAG

No. of catheters used	2.54 ± 0.76 (2–4)
Duration of fluoroscopy (min)	10.2 ± 3.8 (3–24)
The volume of contrast media used (mL)	71.5 ± 45.7 (30–300)

TABLE :3 The mean volume of contrast media used was 71.5 ± 45.7 (range 30–300) mL. In addition to Judkins catheters, Amplatz left 1–2, Amplatz right 2, and hockey stick left/right catheters were among the most commonly used catheters during the procedures. Amplatz left catheters were frequently used in patients with RCAs originating from the left coronary circulation.

DISCUSSION

The coronary anatomical variations depend on the characteristics of the population studied leading to a difference in dominance pattern, presence of IMA, and incidence of CAAs according to different CAG series. The coronary dominance pattern in the present study was 82.6% RCA, 11.5% Co-dominant, and 5.9% LCX, consistent with that found in the literature. [11]The dominance pattern was similar between genders in the present study. IMA was present in 330 (11.0%) patients, without any gender preponderance, less than that reported in a study done by A. Rao et al. [12]The overall incidence of CAA in our study was 2.4% based on Angelini and Khatami's classification. As MB and coronary artery aneurysms are not classified as CAAs in the modified classification, the incidence of CAAs was found to be only 1.1% using the modified classification. Based on the modified classification, 82% of the CAAs were abnormalities in the origin or distribution of a coronary artery (i.e. Group A) and 18% were abnormal fistulae (i.e. Group B). The incidences of CAAs in the present study, according to both classifications, were higher than most of the reported values in the literature. The results of the present study are very similar to those of the Indian study on CAAs, which was conducted by Sidhu NS et al which had an incidence of 3.06%. [13]We observed the separate origin of the LAD artery and LCX from the left SV (i.e. absence of the LMCA) was the most common anomaly (Fig 1). Angiographic incidence of the absence of the LMCA was reported to be 0.43% and 0.18% in two studies performed in India. [14] In the present study, the LMCA was the most common anomalous vessel and the incidence of the absence of the LMCA was 0.8%, slightly higher than the aforementioned studies. [15,16]Coronary arteries may arise from the contralateral coronary sinus or as an early branch of other coronary arteries.

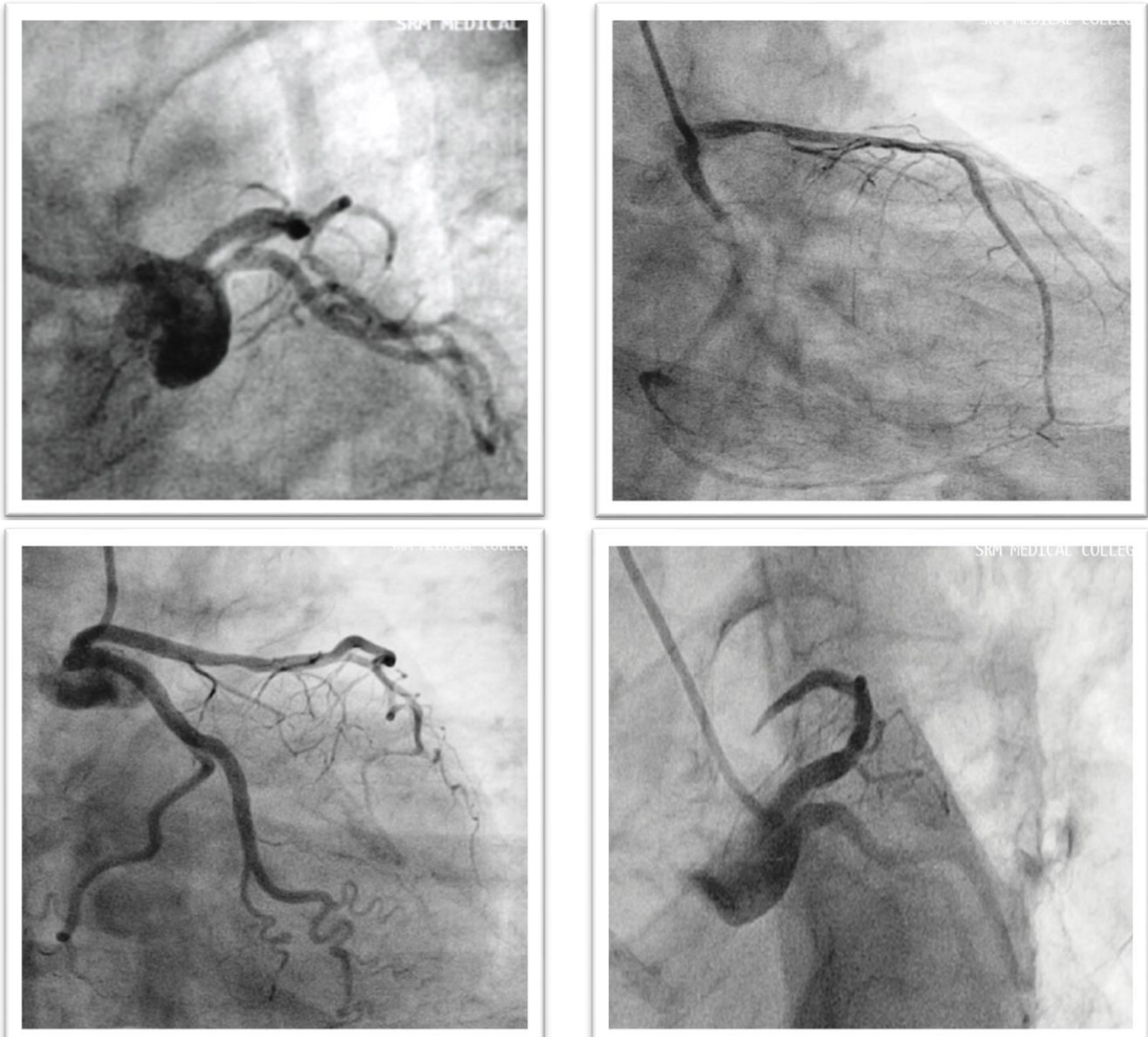


Fig 1: Angiogram shows the separate origin of the left anterior descending (LAD) artery and circumflex artery (CXA) from the left sinus of Valsalva, with the absence of the left main coronary artery

Anomalous origin of the LCX from the right coronary circulation (i.e. the right SV or RCA), which is quite a frequent anomaly, is generally a benign condition. In the present study, four patients had the anomalous origin of the CXA from the right coronary circulation (Fig 2) constituting about 0.13% of the total study population. In a study done by Yogesh Diwan et al, the incidence of anomalous LCX was more common than ours with an incidence of 0.44%. [15]

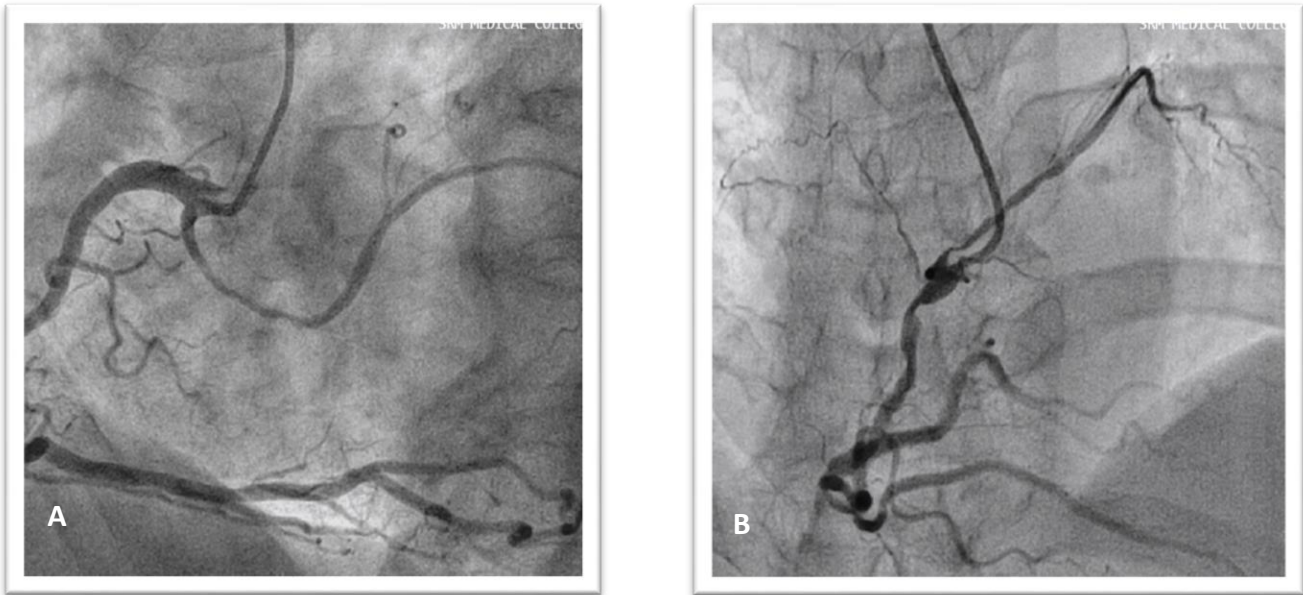


Fig2: Angiogram with anomalous Left circumflex (LCX) originating from Right coronary artery (RCA) in two different patients. Anomalous origin of the RCA from the left coronary circulation is also a common coronary anomaly. The right coronary artery was the commonest anomalous vessel, involved in 19 (0.46%) patients among the total 39 patients who had a coronary anomaly in a study done by N Garg et al in the Indian population.[16] We observed the anomalous origin of RCA in 5 (0.16%) patients with four of them from the left system and one from the aorta directly as shown in Fig 3. Another study done by Sivakumar et al in 1547 patients, observed that anomalous RCA origin was more common than absent left main with an incidence of 0.78% (n=12) which is much higher than our study, emphasizing the fact that the spectrum and incidence of coronary anomalies vary with the different population studied.[17]

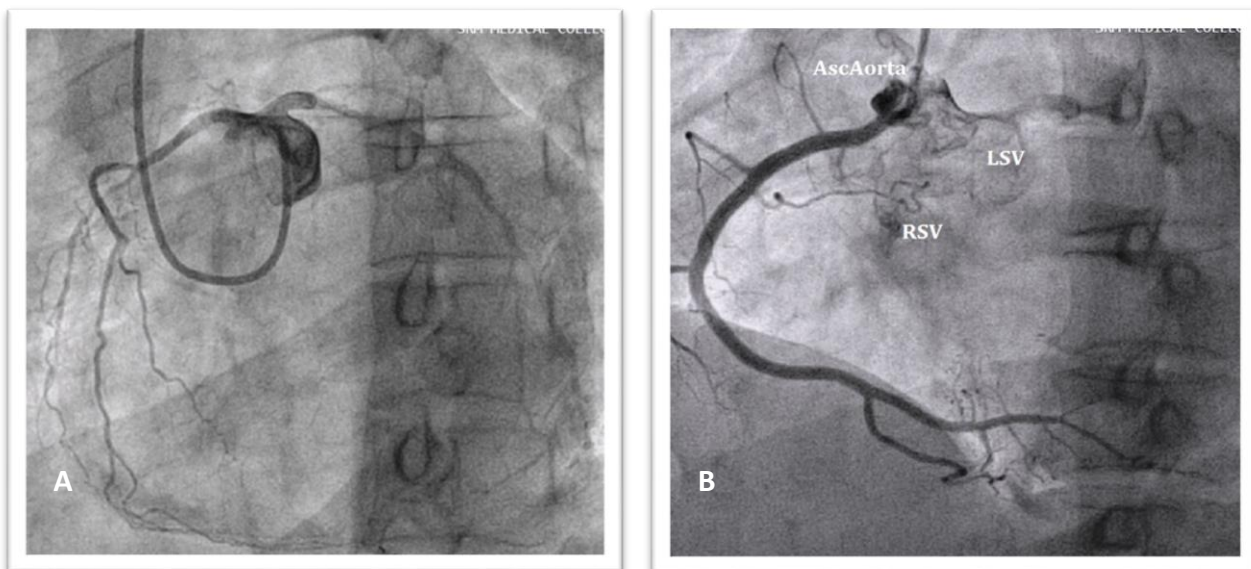


Fig 3: Angiogram shows the anomalous origin of RCA; A-RCA originating from the Left sinus; B-RCA originating directly from Ascending aorta. (LSV-Left sinus of Valsalva; RSV-Right sinus of Valsalva; AscAorta- Ascending Aorta) One of our patients had a single coronary artery arising from a single ostium at the right coronary sinus, branching into a right coronary artery and left coronary artery shortly after the origin (Fig 4). The patient was a post-menopausal female who presented with the acute coronary syndrome with a significant lesion in RCA which was addressed. An aberrant origin of the main stem coronary artery from the right sinus of Valsalva is known as one of the rarest coronary vessel anomalies and the incidence of a single coronary artery is rarer still, reported at 0.024% of the population. Single coronary arteries are usually asymptomatic, but some variants have been reported to carry a risk for serious cardiac events such as sudden

cardiac death and myocardial infarction. The single coronary artery anomaly may be sub-grouped into three categories, according to Lipton's classification. there is no gender predisposition and the management is surgical if presented within 30 years of age with evidence of ischemia and without the atherosclerotic disease. [18]

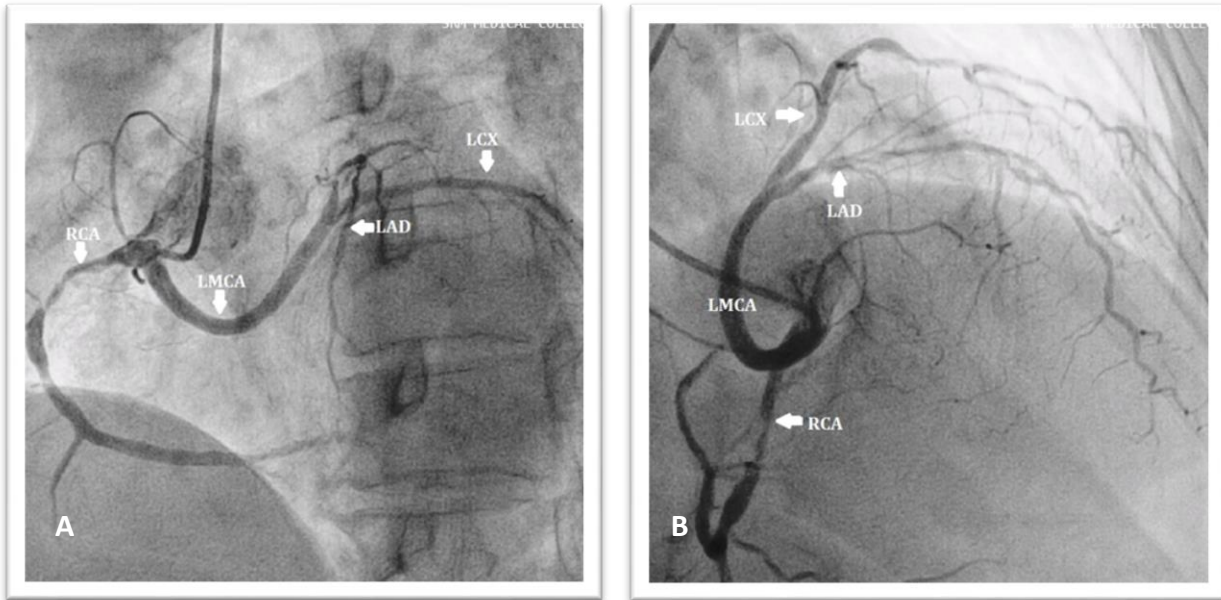


Fig 4: Angiogram shows single main stem originating from the RSV bifurcating shortly after its origin to RCA and LMCA; A- Left Anterior Oblique 40°; B- RAO 30° Cranial 30°

Coronary artery fistula was observed in 6 patients (0.2%) with all draining into RV and was small, nonsignificant fistula without causing any volume overload to cardiac chambers. The prevalence of coronary artery fistulas was thought to be 0.05-0.25% of the population, based on findings in standard coronary angiography studies. However, coronary computed tomography (CT) angiography has been found to detect more coronary artery fistulas,[19] with the prevalence found to be 0.9%. [20]Most of them drain into the right ventricle (40%), right atrium (25%), or coronary sinus, while some drain into the pulmonary artery, left atrium, or left ventricle. [21]In general, the shunt is small and the patients are asymptomatic. However, if the shunt is large, pulmonary hypertension, congestive heart failure, bacterial endocarditis, rupture, and myocardial ischemia may occur. [22]The incidence of MB is estimated to be 1%–5% in the general population based on autopsy findings. The severity of the clinical presentation depends on the extent of systolic compression which has to be assessed during the angiographic procedure. Since most of the coronary flow is in diastole, MB causing systolic compression is not expected to impair myocardial perfusion, ischemia, and angina. It's usually benign if the blockage is less than 50% and symptoms of angina don't appear unless the stenosis is 70% or above.[23]Clinically important MB can cause complications such as vasospasm, angina pectoris, and arrhythmias. Although 30 (1.0%) patients had MB in our study, the incidence was much higher to the tune of 4.77% (n=212) in a study done by Sunil et al in the western Indian population with males affected the most.[24] LAD was the vessel involved in all our cases with mid LAD being the most common site 73%(n=22) of MB. Coronary artery ectasia (CAE) or coronary artery aneurysm is the aneurysmal dilatation of the coronary artery. It is defined as dilatation with a diameter of 1.5 times the adjacent normal coronary artery based on the CASS registry. Its prevalence ranges from 1.2% - 4.9% with a male to female ratio of 3:1. The right coronary artery is most commonly involved followed by LCX and LAD.[25]They are rare anomalies whose aetiologies are unclear but may be associated with inflammatory diseases, connective tissue diseases, and some congenital diseases.[26]In the present study, 162 (5.4%) of our patients had ectasia and 13 (0.4%) had aneurysms. Both of these conditions showed male dominance in the present study, similar to that observed in coronary artery diseases. As both conditions have a similar clinical prognosis, they have to be treated as coronary artery disease. [27]

CONCLUSION

The incidence of CAAs varies, ranging from 0.2% to 8.4% which maybe attributed to the lack of consensus on whether MBs, ectasia, and aneurysms should be classified as CAAs. The relationship between the anomaly and the presenting symptoms is often vague. Fewer than 10% of the patients who experience sudden death due to CAA have symptoms related to the anomaly, with 60%–90% of these patients showing no clinical manifestations. The results of the present study indicate that CAAs are associated with increased radiation and contrast exposure in patients who underwent CAG, owing to

difficulties in ostial cannulation and imaging. The recognition of coronary anatomical patterns, variations, and CAAs is very important, especially when dealing with patients who are scheduled for coronary angioplasty or cardiac surgery. Further prospective studies are required to evaluate the incidence and prognosis of CAAs.

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