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Prevalence and pattern of congenital coronary artery anomalies in adults undergoing coronary angiography at a Tertiary care centre

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Keywords:
coronary artery anomalies, cardiology, congenital anomaly, coronary angiography

ABSTRACT

To study the clinical profile, incidence, and pattern of congenital coronary artery anomalies (CAAs) in adults undergoing coronary angiography. Coronary angiogram of 3600 patients done between September 2017 and September 2021 were reviewed for the presence of CAAs. Angiography data of 3600 patients done between September 2017 to September 2021 was reviewed in the present study and 88 patients were found to have CAAs giving a prevalence of 2.44%. Mean age of patients was found to be 56.86±11.49 years (range 24-75 years). Among these, the anomalous origin and course of the coronaries were the most common anomaly seen in 73 (2.02%) patients, followed by intrinsic anomalies of the coronary arterial system in 13 (0.36%) patients, followed by anomalous coronary termination in 01 patient (0.03%) and single coronary artery 01 patient (0.03%). Overall, the anomalous origin of the right coronary artery (RCA), with RCA arising from left coronary sinus was the most common anomaly seen in 23 patients (0.64%) followed by absence of the left main trunk with a separate origin of the left anterior descending (LAD) and left circumflex artery (LCX) seen in 20 (0.56%) patients. Posterior origin of RCA within right coronary sinus was seen in 13 (0.36%) patients. There were 6 cases of high origin of right coronary artery (RCA) (0.16%). There were 10 patients (0.28%) of LCX arising from right coronary sinus and 01 case of LMCA arising from right coronary sinus. Split RCA was seen in 09 (0.25%) patients whereas split LAD was seen only in 04 (0.11%) patients. Further, there was only 01 (0.03%) patient of anomalous coronary termination (RCA to right atrium fistula) and 01 (0.03%) patient of single coronary artery. The prevalence of congenital coronary anomalies in the study was found to be 2.44%. and commonest anomaly was that of origin and course of the vessels (88%). Though the prevalence of CAAs in this study was similar to that in previous studies, the pattern of anomalies was slightly different from previous studies.

1. Introduction

Coronary artery anomalies (CAAs) are a group of congenital disorders having variable origin, course, termination or some intrinsic anomaly. The incidence of CAAs reported in literature is variable and affects around <1% of general population. It has been reported in 0.3% - 5.6% in different studies in patients undergoing coronary angiography [1], [2]. Although CAAs are rare, in one of the largest autopsy studies conducted to date, the anomalous aortic origin of a coronary artery was the second most common cause of sudden cardiac death (SCD) in young competitive athletes [3], [4].

Different classifications have been used in literature based on functional relevance of each abnormality or anatomical features. Based on functional relevance, CAAs are categorized as (1) anomalies with obligatory ischemia (2) anomalies without ischaemia. In our study CAAs are categorized as (a) anomalies of origin and course, (b) anomalous location of coronary ostium outside normal aortic sinus, (c) anomalies of coronary termination, (d) single coronary artery.

Although CAAs are very rare the precise identification of course and distribution of abnormal coronary vessels is very important before revascularization procedures as they pose difficulty in selective cannulation and to avoid any accidental damage to the vessels. Pattern and prevalence of CAAs has been reported to be affected by geographical variations [5- 7].

In our study, we have presented the angiographic prevalence of CAAs among adult patients under- going coronary angiography at a single tertiary care centre in western India.

2. Materials & Methods

A single-center, retrospective, observational study was carried out at a tertiary care hospital of west- ern India after obtaining the approval of the institutional ethics committee (IEC) of our hospital. Coronary angiogram were independently evaluated by two interventional cardiologists and in case of difference in opinion, a senior cardiologist was consulted to reach the consensus. Coro- nary angiography was done on Philips Allura Centron flat panel digital machine.

Both radial and femoral access were used as per operator preference.

Patients with coronary anomalies such as myocardial bridging, ectasia, and separate origin of the conus artery from the right coronary sinus were excluded from the study.

2.1 Statistical analysis

We used descriptive statistics to study the prevalence of the CAA, where categorical and continuous variables were presented as percentages/frequencies and mean! standard deviation (SD), respectively.

3. Results

A total of 3600 angiograms done in a span of 4 years were included in the study which yielded a total of 88 cases (2.44%) of different forms of CAAs. Among these, there were 56 (63.64%) males and 32 (36.36%) females. The mean age of the patients with CAAs was 56.86±11.49 (range 24-86) years. The highest number of patients with anomalous coronaries, 26 (29.55%) were seen in the age group of 41- 50 years followed by 25 (28.41%) in 51-60 years, 19 (21.60%) in 61-70 years, 12 (13.66%) in 71-80 years, 02 (2.21%) in 21-30 years, 03 (3.41%) in 31-40 years and 01 patient in 81-90 (1.34%) years age group. The most common indication for undergoing angiography among these patients was acute coronary syndrome (N=54, 61.36%), followed by chronic stable angina (CSA) (N=28, 31.81%), valvular heart disease (N=05, 5.68%), and dilated cardiomyopathy (N=01, 1.14%) (Table 1).

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3.1 Anomalies of origin and course

Absence of Left Main Trunk with Separate Origin of left anterior descending artery (LAD) and left circumflex artery (LCX): The separate origin of LAD and LCX from the left coronary sinus with the absence of the left main trunk was seen in 20 patients having an angiographic prevalence of 0.56% and anomaly incidence of 22.72%. This was the second most common anomaly observed in our study.

3.2 Anomalous location of coronary ostium outside normal aortic sinus

This was the most common anomaly with an angiographic prevalence of 1.47 % and anomaly incidence of 60.23%. Out of these, 23 patients had RCA arising from left coronary sinus with an angiographic prevalence of 0.64% and anomaly incidence of 26.14%.

Whereas posterior origin of RCA within right coronary sinus was seen in 13 patients with an angiographic prevalence of 0.36% and anomaly incidence of 14.77%, further there were 6 patients with high origin of RCA with an angiographic prevalence of 0.16% and anomaly incidence of 6.82%.

Anomalous origin of LCX from right sinus: It was observed in 10 patients with an angiographic prevalence of 0.28% and anomaly incidence of 11.36%.

Anomalous origin of LMCA from right sinus: It was observed only in 1 patient with an angiographic prevalence of 0.03% and anomaly incidence of 1.14%.

3.2.1 Anomalies of intrinsic coronary artery anatomy

Split RCA: Split RCA was seen in 09 patients with an angiographic prevalence of 0.25% and anomaly incidence of 10.23%.

Split LAD: Split LAD was seen in 04 patients with an angiographic prevalence of 0.11% and anomaly incidence of 5.69%.

3.3 Anomalous coronary termination

Only 01 patient of anomalous coronary termination; RCA to right atrium fistula was seen with an angiographic prevalence of 0.03% and anomaly incidence of 1.14%.

3.4 Single coronary artery

Single coronary artery arising from left coronary cusp was reported with angiographic prevalence of 0.03% and anomaly incidence of 1.14%.

4. DISCUSSION

CAAs are diverse group of rare congenital disorders with abnormal origin, course, termination and intrinsic anomaly. CAAs have been categorized mainly based on functional relevance or anatomical features. In our present study, the overall prevalence of CAAs has been reported to be 2.44%, which is in agreement with 0.3%-5.64% incidence reported by various other studies [8], [9]. The prevalence of anomalies was higher in males with a male to female ratio of 1.75:1. This is in tune with other studies which have reported male predominance of prevalence upto the tune of 3:1. This high number may be attributed to the fact that fewer number of females undergo angiography as compared to males, in our country.

The most common anomaly seen in this study was origin of right coronary artery (RCA) from left coronary sinus. It was reported in 23 patients with an angiographic prevalence was 0.64%. It was similar to to our

results, its high prevalence has been reported by (0.22%) [9] and (0.40%) [17]. It was reported to be most common anomaly by (0.37%) [18] and second most common anomaly by [17]. Whereas, in contrast to our study it was reported by with prevalence of 0.10% [11] with prevalence of 0.1% [12]. In this anomaly, the RCA originates from an orifice located anterior to the left main ostium in the LCS and courses either anterior to pulmonary artery, or between aorta and pulmonary artery (inter-arterial) or posterior to aorta (retro-aortic) of which inter-arterial course is potentially serious anomaly and may result in myocardial ischemia and life-threatening clinical manifestations including myocardial infarction, ventricular tachycardia, syncope and sudden death in the absence of significant atherosclerosis [10].

The second most common anomaly observed in the study was absence of the left main trunk with separate origin of the left anterior descending (LAD) artery and left circumflex artery (LCX). This was observed in 20 patients with an anomaly incidence of 22.72% and angiographic prevalence of 0.56%. Other studies have suggested its prevalence to be 0.74% [9], 0.37% [10], 0.69% [11], 0.16% [12], 0.45% [13], 0.43% [14, 15] 0.6% [16]. This variable of CAA is clinically benign.

Posterior origin of RCA within right coronary sinus was seen in 13 (0.36%) patients. Its prevalence has been reported to be 0.16% by [13] which is potentially benign. There were 6 cases of high origin of right coronary artery (RCA) (0.16%)

Next CAA was LCX arising from right coronary sinus (RCS) or from proximal RCA. In our study, LCX arising from RCS was reported to be the fourth most common anomaly present in 10 patients with a prevalence of 0.28%. Similar incidence of this anomaly has been reported by (0.28%) and (0.25%). However, it is less than that reported by (0.34%) [18], and (0.33%) [13] and more than that reported by (0.17%) [9]. This anomaly is usually benign, however, carries the risk of accidental ligation or injury during valve surgery.

Anomalous origin of left main coronary artery from right coronary sinus was found only in 1 patient with an angiographic incidence of 0.03%. This is a rare anomaly with an estimated incidence of 0.03- 0.15% in various studies [17], [19]. However, this anomaly has great clinical significance be- cause of its association with sudden cardiac death, especially during vigorous exercise [2], [19].

Split RCA defined as the one with early PDA (posterior descending artery) branch was the fifth most common anomaly in our study. It was seen in 9 patients with an angiographic prevalence of 0.25% which was reported to be most prevalent coronary anomaly (0.84%) by [16]. Split LAD defined as the one with a major septal or major diagonal branch further giving small septal branches along with LAD was seen only in 4 (0.11%) patients.

In our study, 1 case with anomalous coronary termination was seen (right coronary artery to right atrium fistula) with a prevalence of 0.03%, and 1 case of single coronary artery from left coronary sinus was found with prevalence of 0.03% which was almost similar to [13] (0.02%) Anomalous origin of the RCA from the left coronary sinus is an important anomaly with a reported incidence of 0.1%. Because this anomaly involves a course between the aorta and pulmonary artery, it has been implicated in sudden cardiac death, and some recent studies have reported a prevalence of 30% among asymptomatic adolescents who have suffered sudden cardiac death [20], [21], [24].

Anomalous origin of LCX has two possibilities: (1) separate LCX from the left coronary sinus or (2) LCX arising from the RCA or from the right coronary sinus near the RCA ostium. In the latter situation, the

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course is usually retroaortic and clinically benign [21], [23].

Anomalous origin of the LMCA from right coronary sinus is quite rare, with an estimated incidence of 0.03-0.15%. In this case, it may take four possible courses [25]:

(a) Interarterial course: Between the aortic root and the pulmonary artery. The LMCA initially courses up and then behind the right ventricular outflow tract before travelling anteriorly to its normal point of bifurcation.

(b) Septal course. The LMCA may course intramyocardially or subendocardially, along the floor of the right ventricular outflow tract. It will then surface in the midseptum, at which point it branches into the LAD (only the mid and distal segments are present) and LCX (the initial portion courses toward the aorta). Another clue is the presence of the unusual septal perforator branches arising from the LMCA

(c) Anterior (to the right ventricular outflow tract) course. The LMCA passes anteriorly over the right ventricular outflow tract, making a cranial anterior loop in which the artery passes initially rightward then up and over the right ventricular infundibular free wall before reaching the inter-ventricular groove and then dividing normally.

(d) Retroaortic (posterior to the aortic root) course. The LMCA may arise to the right of the RCA and pass posterior to the aortic root.

This anomaly is clinically significant, as it is associated with sudden cardiac death, especially during vigorous exercise [26]. The potential lethality of this anomaly is due to the course followed by the LMCA on arising from the right coronary sinus.

There are a number of postulated mechanisms to explain the mechanisms of ischaemia associated with LMCA anomalies. The normal coronary artery arises at a perpendicular angle to the aorta, in contrast to the anomalous coronary artery arising from the opposite coronary sinus, which comes off at an acute angle and bends over itself to reach its normal supply territory. Because of this, the ostium of the anomalous coronary artery is smaller, with a slit-like ostium, as compared with the normal circular ostium. The magnitude of ischaemic risk is related to the degree of angulation.

These complications commonly occur during or immediately after exercise. Exercise leads to expansion of the aortic root and pulmonary trunk, stretching the aortic wall and leading to compression and exaggeration of the slit-like ostium causing further ischaemia [27], [28].

Another theory refers to the initial course of the CAA. When the anomalous artery course is interarterial, between the aorta and pulmonary artery, the increase in the pressure in the two vessels occur- ring during exertion would produce a compression of the vessel, myocardial ischaemia and carries a high risk for sudden cardiac death in both adults and teenagers [29]. If the initial pathway is an intramural course, that segment of the coronary artery may be compressed and deformed within the aortic wall during periods of hypertension [24], [30].

With respect to RCA anomalies, the acute angulation at the coronary takeoff may increase or be- come kinked during exertion; the slit-like orifice may become compressed by exercise-induced aortic dilatation; the intramural segment may be compressed by the aortic valve commissure; and the presence of an ostial

ridge functioning as a #valve$ which restricts flow during exertion [31], [32].

Angelini has demonstrated with intravascular ultrasound lateral luminal compression of the intra- mural portion of the coronary artery and compression of the coronary artery between the aorta and the pulmonary artery [22], [31].

Some authors suggest that anomalous pathways may result in endothelial damage and dysfunction leading to coronary artery spasm contributing with other mechanisms (mentioned above) to cause myocardial ischaemia [33].

Most CAAs are asymptomatic and are detected incidentally during angiography. Some anomalies present as sudden cardiac death in young subjects. [34] reported that 19% of deaths in athletes are attributable to CAA.

When symptomatic, modes of presentation include episodic chest pain, syncope, dyspnoea, heart failure, ventricular arrhythmias, myocardial infarction and sudden death. Symptoms typically occur after heavy exercise and are usually associated with an anomalous origin of the LMCA or the RCA from the opposite coronary sinus.

Limitations
This was a Single Centre study and does not represent the true prevalence of CAAs in general population. Further, patients who underwent coronary angiography for acute chest pain, acute coronary syndrome were included in the study, which further does not represent the general population.

5. CONCLUSION

The prevalence of CAAs in the present study was 2.44%. Among these, the most common were the anomalies of origin and course. However, patterns of anomalies reported in the present study were different from the similar studies done in other parts of the world. Overall, the anomalous location of coronary ostium outside normal aortic sinus of Valsalva was the most common anomaly followed by absence of the left main trunk with separate origin of LAD and LCX. Although CAAs are rare, their accurate recognition is very import because of their clinical significance and appropriate management of cardiac patients. Though most of the CAAs were benign, their angiographic recognition is important for further cardiac interventions in the form of coronary angioplasty or surgery.

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Table 1: Baseline characteristics and pattern of coronary anomalies. All values are presented as mean ± SD or number (%). CAA: coronary artery anomalies.

CHARACTERISTICS	**NUMBERS**
Total number of coronary angiograms reviewed	3600
Number of patients with CAAs	88 (2.44%)
Anomalies of coronary origin and course	73 (2.02%)
Anomalies of coronary arterial anatomy	13 (0.36%)
Anomalies of coronary termination	1 (0.03%)
Single coronary artery	1 (0.03%)
Mean age (range) in years of patients with CAAs	56.86 +/- 11.49 years
GENDER DISTRIBUTION OF PATIENTS WITH CAAs	
Number of males	56 (63.64%)
Number of females	32 (36.36%)
INDICATION FOR CORONARY ANGIOGRAPHY	
Acute coronary syndrome	54 (61.36%)
Chronic stable angina (CSA)	28 (31.81%)
Valvular heart disease	05 (5.68%)
Dilated cardiomyopathy	01 (1.14%)

TABLE 2: Prevalence of various coronary artery anomalies. LAD: left anterior descending, LCS: left coronary sinus, LCX: left coronary circumflex, LCA: left coronary artery, RCA: right coronary artery, RCS: right coronary sinus.

CORONARY ANOMALY	NO. OF PA-TIENTS (n=3600)	ANGIOGRAPHIC INCIDENCE (%)	ANOMALY INCI-DENCE (%)
1) ANOMALOUS ORIGIN AND COURSE			

A) ABSENT LEFT MAIN TRUNK			
> Separate origin of LAD & LCX	20	0.56	22.72
B) ANOMALOUS LOCATION OF CORONARY OSTIUM OUTSIDE NORMAL AORTIC SINUS			
RCA arising from left coronary sinus	23	0.64	26.14
Posterior origin of RCA within right coro-nary sinus	13	0.36	14.77
High origin of RCA	6	0.16	6.82
LCX arising from right coronary sinus	10	0.28	11.36
LMCA arising from right coronary sinus	1	0.03	1.14
2) ANOMALIES OF INTRINSIC CORONARY ARTERY ANATOMY			
Split RCA	9	0.25	10.23
Split LAD	4	0.11	5.69
3) ANOMALIES OF CORONARY TERMINATION			
RCA to right atrium fistula	1	0.03	1.14
4**) SINGLE CORONARY ARTERY**	1	0.03	1.14
TOTAL	88	2.44	

Figure 1:- Coronary angiogram showing posterior origin of RCA in right coronary sinus.

Figure 1: Coronary angiogram showing posterior origin of RCA in right coronary sinus

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Figure 2:- Coronary angiogram showing anomalous RCA and LCXarising from Right coronary sinus.

Figure 2: Coronary angiogram showing anomalous RCA and LCX arising from Right coronary sinus

Figure 3:- Coronary angiogram showing anomalous origin of RCA from Left Coronary Sinus.

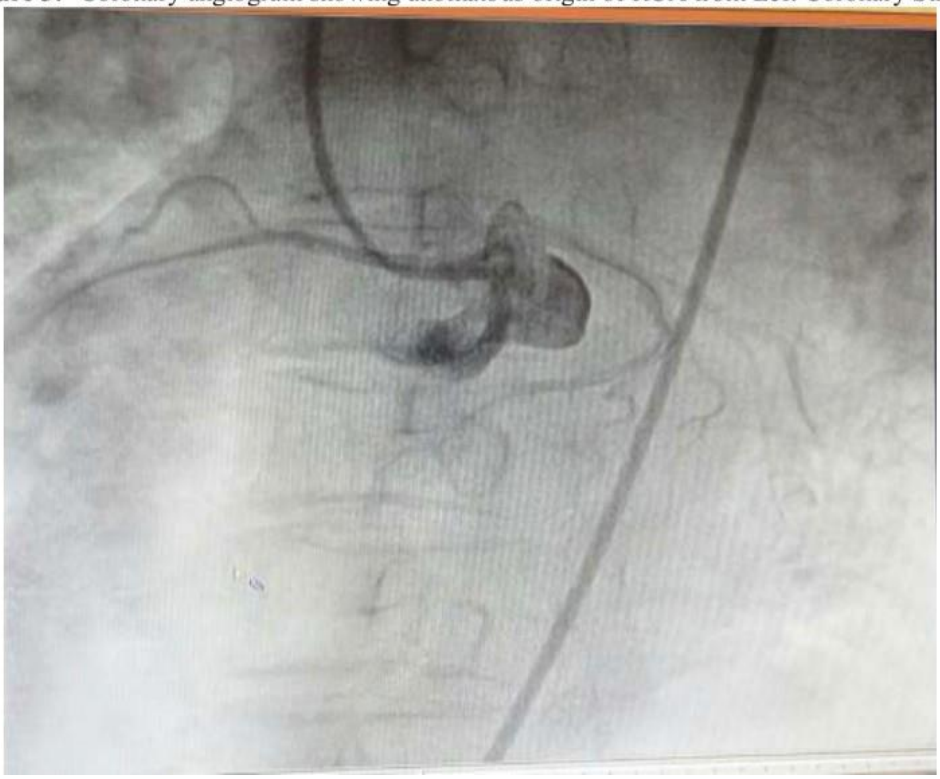

Figure 3: Coronary angiogram showing anomalous origin of RCA from Left Coronary Sinus

Figure 4:- Coronary angiogram showing LCX arising from Right Sinus.

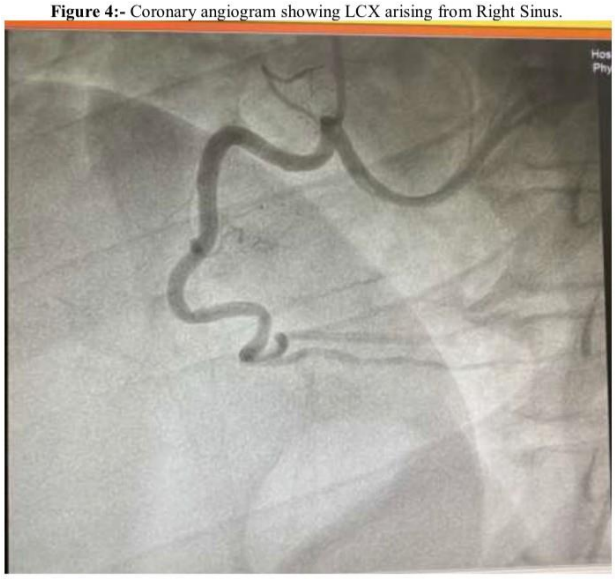

Figure 4: Coronary angiogram showing LCX arising from Right Sinus

Figure 5:- Coronary angiogram showing anomalous origin of Left Main Coronary Artery from Right coronary sinus.

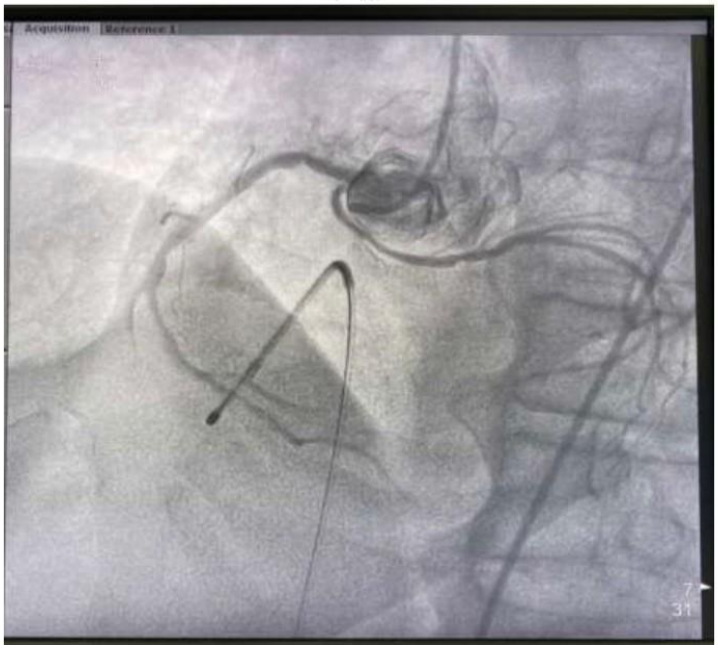

Figure 5: Coronary angiogram showing anomalous origin of Left Main Coronary Artery from Right coronary sinus

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Figure 6:- Coronary angiogram showing Split Left Anterior Descending Artery.

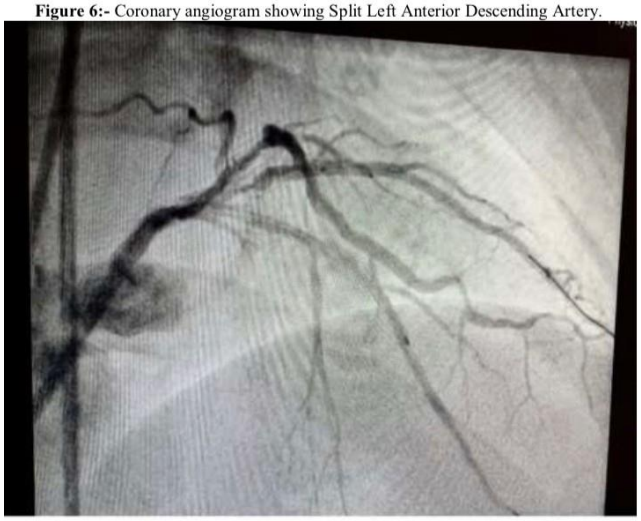

Figure 6: Coronary angiogram showing Split Left Anterior Descending Artery

Figure 7:- Coronary angiogram showing Split Right coronary artery.

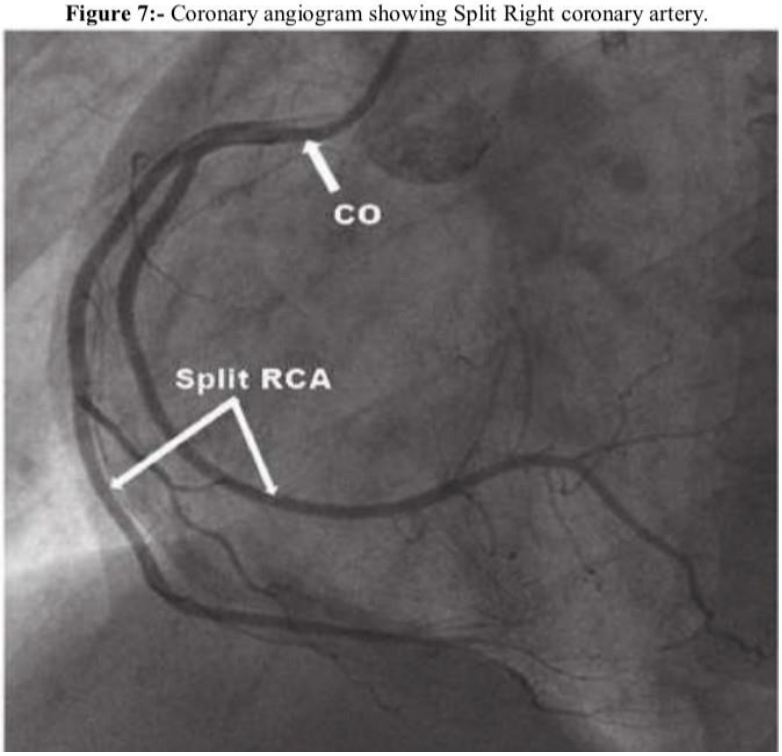

Figure 7: Coronary angiogram showing Split Right coronary artery

Figure 8:- Single coronary artery from left sinus.

Figure 8 - Single coronary artery from left sinus

Figure 9:- Coronary angiogram showing right coronary artery to right atrium fistula.

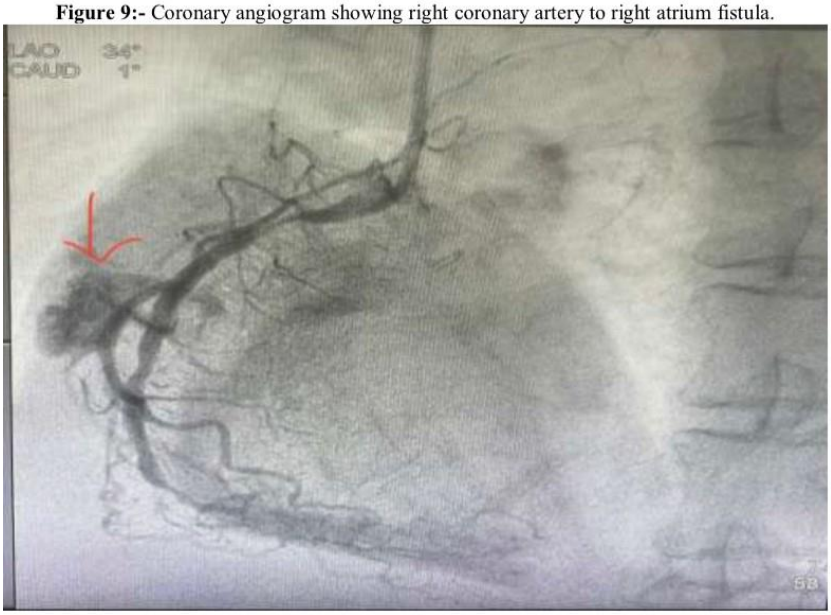

Figure 9 - Coronary angiogram showing right coronary artery to right atrium fistula

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Figure 10:- High origin of RCA.

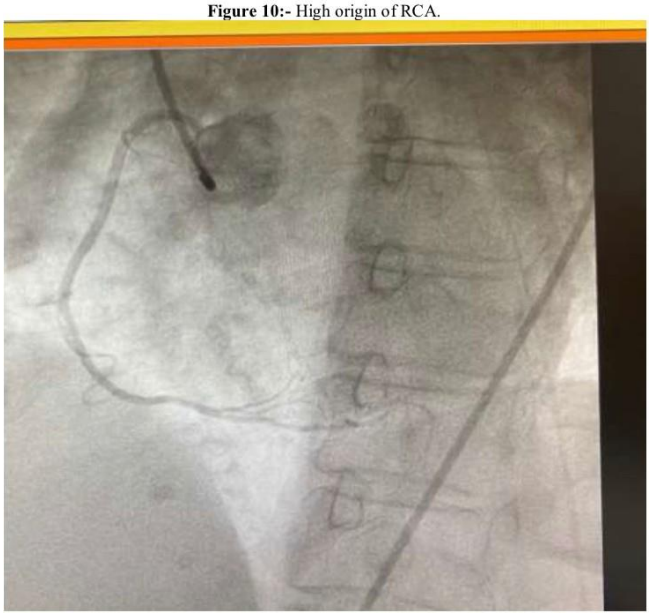

Figure 10: High origin of RCA