

Prevalence and Patterns of Coronary Artery Anomalies in a Tertiary Cardiac Center of Nepal: A Coronary Angiographic Study

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ABSTRACT

Introduction

The prevalence of coronary artery anomalies (CAAs), though rare, is nonetheless globally found in 0.2% to 1.3% of patients undergoing coronary angiography (CAG). We aimed to investigate the prevalence and patterns of CAAs.

Methods

The data was collected retrospectively by analyzing the coronary angiographic data of 6000 consecutive adult patients undergoing coronary angiography between January 2016 to December 2020 at the Manmohan Cardiothoracic Vascular and Transplant Center.

Results

Among 6000 patients, 89 (1.4%) had CAAs, with anomalies in the right coronary artery (RCA) being the most common (73%). The presence of a dominant RCA was found in 48 (53.9%) patients. The anomalous origin of RCA arising from the left sinus of valsalva was present in 33 (37.1%) patients. The left main (LM) anomaly was present in 49 (55%) patients with absent LM in 29 (32.6%) and a short LM in 20 (22.5%) patients, respectively. The left anterior descending (LAD) artery anomalies were present in 42 (47.2%) patients - out of which 29 (32.6%) had a separate origin of LAD from the left coronary sinus and 13 (14.6%) had a dual LAD. The left circumflex artery (LCx) anomalies were evident in 32 (35.9%) patients including separate origin from left coronary sinus in 29 (32.6%) and anomalous origin from the right coronary sinus in 3 (3.4%) patients, respectively. Two patients (2.2%) had coronary artery fistulas, both arising from LAD.

Conclusion

Though rare, our study did discover the prevalence of CAAs to be 1.4%.

Keywords

Coronary angiogram, coronary artery anomaly, prevalence

INTRODUCTION

The anatomical variations of the coronary artery that are present in a small percentage of population are known as coronary artery anomalies (CAAs)¹ which are generally encountered incidentally during the coronary angiography (CAG) or at autopsy.² The three basic elements for development of coronary vasculature are the sinusoids, the in situ vascular endothelial network, and coronary buds on the aortic sinuses. Any abnormalities in the development of the coronary buds or the endothelial network are responsible for CAAs and the persistence of sinusoids results in coronary fistulae.³

The coronary arteries arise from the aortic sinuses, converging towards the apex of the heart. Normally, there are three main coronary arteries, the right coronary artery (RCA), left circumflex artery (LCx) and left anterior descending (LAD), with the LCx and LAD arteries arising from a common stem, the left main coronary artery (LMCA). The LMCA originates from the left aortic sinus just below the sinotubular ridge of the aorta and passes between the main pulmonary artery and the left auricle and finally bifurcates into LAD and LCx arteries.⁴ In around one-third of cases, the LMCA trifurcates to produce a ramus intermedius (RI) branch. The LAD arises from the LMCA bifurcation or separate ostia if LMCA is absent. It runs on the anterior interventricular groove towards the apex. The LCx arises from the LMCA bifurcation or separate ostia if LMCA is absent and courses within the left atrioventricular groove towards the inferior interventricular groove. The RCA arises from the right aortic sinus inferior to the origin of left coronary artery, passes anteriorly and to the right between the right auricle and the pulmonary artery and then descends vertically in the right atrioventricular sulcus.⁵

The prevalence of CAAs in the angiographic and autopsy series is 0.6% to 1.3% and 0.3% respectively and about 80% of them are benign while 20% are known to produce life threatening symptoms like arrhythmias, syncope, myocardial infarction, and sudden cardiac death.⁶ Coronary artery anomalies are the second most common cause of sudden cardiac death (SCD) in young athletes.⁷

Data reporting the patterns of CAAs is scarce in Nepal. Hence, this study was designed to compute the prevalence and contemporary patterns of CAAs in patients undergoing coronary angiography in the Nepalese population at a large tertiary cardiac care center.

METHODS

This was a single-centered retrospective study in which the coronary angiograms performed

in 6000 consecutive patients between January 2016 to December 2020 were retrieved from the archives of the Manmohan Cardiothoracic Vascular and Transplant Center's cardiac catheterization laboratories and were later prospectively reviewed by the two independent interventional cardiologists of the same center in the PHILIPS XCELERA Viewer before being finally classified. The indications for performing CAG were chest pain, angina equivalent symptoms, evaluation of syncope, evaluation of cardiomyopathy, and pre-operative evaluation for a non-cardiac surgery. Coronary anomalies in patients undergoing catheterization for congenital heart disease were excluded. Because of a lack of clarity regarding the definition of myocardial bridges,^{8,9} we also excluded them in the category of coronary anomalies. There was no inter-observer variability between the investigators.

This study was conducted after obtaining the ethical clearance from the Institutional Review Committee (IRC) of Institute of Medicine [Reference number: 38/ (6-11) E2077/078]. Also, all patients have given their informed consent prior to their inclusion in the study.

Coronary angiograms were performed through standard transradial or transfemoral techniques. 1% Lidocaine was injected into the superficial skin as a local anesthesia. The modified Allen's test was performed prior to CAG for the radial procedure. The patient was comfortably positioned in the catheterization laboratory. After draping the arm, the radial artery was cannulated using a 20 Gauge needle 1 to 2 cm proximal to the bony prominence of distal radius at a 30-to-45 degree angulation. A radial wire of 0.018 inch was carefully introduced into the vascular lumen through the IV cannula. A 5-French radial sheath (5F) was advanced over the wire into the artery. Finally, a radial cocktail comprising of unfractionated heparin 2500 U, Verapamil 2.5mg, and Nitroglycerin 50 to 100 mcg were administered to prevent radial artery occlusion and spasm. The selective cannulation of aberrant arteries were difficult and time-consuming and we used Tiger, Judkins left and right, left Amplatz, and Extra back up guide catheters for angiography and angioplasty. A right femoral approach was used if the radial artery was too small or too circuitous for catheters to manipulate. A low osmolar contrast media (Iohexol) was used in all cases to make angiographic images at different angles.

Coronary dominance was defined as the artery which gave off the posterior descending artery (PDA) and posterolateral ventricular (PLV) branches. The term co-dominant was used if PDA originated from RCA and PLV originated from LCx. Coronary artery anomalies were recognized according to the classification proposed by Angelini et al¹⁰ which divides coronary artery anomalies into three

categories: anomalies of origination and course, anomalies of intrinsic coronary arterial anatomy, and anomalies of coronary termination.

The normal length of left main coronary artery (LMCA) was defined as 10-15 mm. LMCA was labeled short if it was less than 5 mm and long if it was more than 15 mm. If the LMCA trifurcated to give an intermediate branch, it was labeled as ramus intermedius (RI) artery. The term "dual LAD" was used if two branches supplied the usual distribution of LAD. A coronary anomaly with a high takeoff or ectopic origin referred to a left or right coronary ostium, which arose more than 5 mm above the sinotubular junction rather than at the aortic sinus.¹¹ The term coronary artery aneurysm was defined as the condition in which coronary artery segments had a diameter more than 1.5 times greater than that of the diameter of the normal adjacent coronary segments and involved less than 50% of the total length of the vessel while the term "ectasia" was used to describe a condition in which diffuse dilatation of coronary arteries occurred over more than 1.5 times than the normal diameter that involved 50% or more of the length of the artery.¹² Coronary artery fistulas were referred to the abnormal connection between the coronary artery and other structures, including the coronary vein, heart chamber, or pulmonary artery.

RESULTS

The coronary artery anomalies were seen in 89 patients (prevalence of 1.4%). The mean age of the subject cohorts was 55.6±12.8 years. Among 89 patients, 65 were males (73%) and 24 were females (27%).

Overall, the most commonly used catheter for successful cannulation of the anomalous coronaries was Tiger (58/89, 65.2 %). In case of patients with RCA anomalies (n = 65), the Judkins left (JL) was found out to be the most successful catheter (19/65, 29.2 %). The median (IQR) contrast volume was 45 ml (37.5 – 100 ml). The maximum volume of contrast given was 250 ml. Similarly, the median (IQR) fluoroscopy time was 8 min (5.5 – 11.5 min).

CAG was done through right radial artery in 67 (75.3%) patients, right femoral artery in 17 (19.1%) patients, and right radial approach was shifted to right femoral approach in 5 (5.6%) patients due to occurrence of radial artery spasm during procedure.

Coronary artery circulation was right dominant in 48 (53.9%) patients, left dominant in 38 (42.7%) patients, and co-dominant in 3 (3.4%) patients. The left main coronary artery showed trifurcation into ramus intermediate branch in 6 (6.7%) patients. Among 89 subject cohorts, the most common anomalous coronary vessel was found out to be right coronary artery (RCA) in 65 (73%) patients.

Table 1. Distribution of coronary anomalies

Characteristics	Number (%)
Approach	
Femoral	17 (19.1)
Radial	67 (75.3)
Radial converted to femoral	5 (5.6)
Dominance	
Right	48 (53.9)
Left	38 (42.7)
Codominant	3 (3.4)
LM anomaly	49 (55.0)
Absent	29 (32.6)
Short	20 (22.5)
LAD anomaly	42 (47.2)
Separate origin from LCS	29 (32.6)
Dual LAD	13 (14.6)
Ramus intermedius (RI)	6 (6.7)
LCX anomaly	32 (35.9)
Separate origin from LCS	29 (32.6)
Separate origin from RCS	3 (3.4)
RCA anomaly	65 (73.0)
Separate origin from LCS	33 (37.1)
Separate origin of right conal artery and RCA	12 (13.5)
High up	7 (7.9)
Anterior and downward pointing	13 (14.6)
Coronary cameral fistula	2 (2.2)
LAD to RA	1 (1.1)
LAD to LA	1 (1.1)

The RCA arising anomalously from the left sinus of valsalva was present in 33 (37.1%) patients. The separate origin of RCA and conus artery was seen in 12 (13.5%) patients. The ectopic origin of RCA in the form of high take off was present in 7 (7.9%) and the anterior and downward pointing course was found in 13 (14.6%) patients. The left main (LM) anomaly was present in 49 (55%) patients with absent LM in 29 (32.6%) and a short LM in 20 (22.5%) patients, respectively. The left anterior descending (LAD) anomalies were present in 42 (47.2%) patients; out of which 29 (32.6%) had a separate origin of LAD from the left coronary sinus and 13 (14.6%) patients had a dual LAD. Similarly, the left circumflex artery (LCx) anomalies were evident in 32 (35.9%) patients which included separate origin from left coronary sinus in 29 (32.6%) and separate origin from the right coronary sinus in 3 (3.4%) patients, respectively. There were 2 patients (2.2%) with coronary artery fistulas (CAFs) and they mainly occurred in the LAD. One CAF entered on the right atrium while the other entered on the right atrium. Table 1 elicits the details of distribution of all coronary anomalies that were found in our study.

DISCUSSION

CAAs were described in the 18th century, but the first scientific statement on their relevance and comprehensive classification was first published in 1969¹³ and was later revised in 2000.¹⁴ Due to a well-known fact that the most coronary artery anomalies are asymptomatic, they are usually encountered coincidentally during routine coronary angiography or at autopsy.⁶ The prevalence of coronary artery anomalies in our study was 1.4%. Likewise, the prevalence of CAAs was 1.3% as shown by the largest study ($n = 26,595$) conducted by the Cleveland Clinic Foundation in North America in 1990.¹⁵

Our study showed a dominant right coronary system in 53.9% of patients – a finding which is similar to that described in the earlier literature in which 60.58% patients had a right dominant coronary circulation.¹⁶ The origin of the posterior descending artery from either the right (70%) or the left (10%) coronary artery defines the coronary dominance (codominance in 20% cases), with the dominant artery usually providing blood supply to the sinoatrial and atrioventricular nodes.¹⁴

The most frequent coronary anomaly reported in an earlier study was the origin of RCA from LSV.¹⁵ In a single-centered study conducted in Korea, the most common anomaly (39.8%) was an anomalous origin of RCA from the LSV.¹⁵ In our study too, the origin of RCA from LSV was the most common coronary anomaly (37.1%). This signifies a clinically significant anomaly because the inter-arterial course between the aorta and the main pulmonary artery may lead to myocardial ischemia and sudden death due to compressive phenomenon. There are four patterns of anomalous origin of coronary arteries while they cross over to the opposite sinus, i.e. 1. A course anterior to the pulmonary trunk or the right ventricular outflow tract, 2. An inter-arterial course between the pulmonary artery and the aorta, 3. A septal course through the interventricular septum, and 4. A retro-aortic course posteriorly between aortic root and the left atrium.¹⁷ Of these, the inter-arterial course is clinically malignant because it is strongly associated with myocardial ischemia and sudden death. The assumed mechanism of sudden death is ostial closure between the aorta and pulmonary artery and the squeezing of the ostium during exercise, with sudden interference in the coronary blood flow.¹⁶ The conus artery is generally considered to be the first branch of the RCA in the right sinus of valsalva (RSV). Both, the right ventricular outflow tract and the anterior free wall of right ventricle are supplied by the conus artery. Sometimes, it forms an anastomosis with the corresponding branch of left coronary artery known as Vieussens arterial ring.¹⁸ In about 50% of humans, the conus artery originates from a

discrete ostium in the RSV. In contrast, our study reported that 13.5% of patients had a separate conus artery arising from the RSV. Similarly, it is a rare phenomenon to encounter a high take-off of the RCA (incidence of 0.019% to 0.17%)¹⁹ which do not arise from the RSV that may eventually provoke myocardial ischemia and sudden death during exertion due to decreased coronary perfusion. The long course of the RCA originating from the tubular portion of the aorta creates an acute angle at the origin. In addition, the ostium may sometimes have a slit-like shape. Both anomalies are clinically significant and may impair myocardial perfusion.²⁰ In our case, the RCA originated from the anterior surface of the ascending aorta high above the aortic sinus in 7.9% of patients.

The abnormal origin of coronary artery is yet another rare form of CAA whose prevalence differs among different countries. The major abnormal origin of coronary artery is the RCA in India and China whose prevalence varies from 0.38% to 1.3%²¹ while in Europe, the major abnormal origin is the left coronary artery with a frequency of 0.4% to 3.3%.²² Such coronary anomaly can lead to death during infancy with the most notorious anomaly being anomalous origin of left coronary artery from the pulmonary artery (ALCAPA).²³ In our present study, we did not find any severe abnormal origin of the coronary artery, which might be ascribed to the fact that most patients in our study were adults.

The LMCA originates from the left coronary sinus and passes leftward, posterior to the pulmonary trunk, and finally bifurcates into LAD and LCx arteries.²⁴ The left main artery is absent in 0.41% of cases in which the LAD and LCx arteries bifurcate separately and represents 30.4% of all coronary anomalies.²⁵ Even in our study, LM was absent in 32.6% of patients with separate origins of LAD and LCx arteries for the same proportion. The rate of development of atherosclerosis in LAD and LCx branches is influenced by the length of LMCA with earlier development of atherosclerosis in patients with short LMCA. The length of LMCA could also be considered as a hereditary factor for the development of coronary artery disease. The mechanisms involved seem likely to be related to modification of mechanical and hydraulic factors resulting from difference in the length of the left coronary artery trunk.²⁶ In our study, 22.5% of patients had short LMCA. Studies regarding proportion of short LM are scarce.

The LAD has the most constant course among all coronary arteries. Duplication of the LAD is a rare anomaly.²⁷ The classification of dual LAD was first described by Spindola-Franco et al²⁸ in 1983 which consisted of a short LAD that ends high in the anterior interventricular groove and a long LAD that most commonly originates as an early branch

of the LAD proper (types 1-3) and rarely originates anomalously from the RCA (type 4). Dual LAD was found in 13 (14.6%) patients in our study, out of which 9 patients had type 1, 3 patients had type 2, and 1 patient had type 4 variants, respectively. The prevalence of dual LAD in their largest case series was found out to be 1% (23 cases in 2140 patients).²⁷

The origin of the LCx from the right sinus of Valsalva is a well-known anatomical variation. Although difficulties may be encountered during diagnostic procedure, recognition and adequate visualization of the anomaly is essential in patients undergoing PCI, coronary artery surgery or prosthetic valve replacement.²⁹ An avascular area in the posterior lateral left ventricular myocardium during selective opacification of the left coronary artery suggests that there is an anomalous origin of the LCx.³⁰ The anomalous origin of the LCx from the right sinus of Valsalva was seen in 3 (3.4%) patients in our study. However, this anomaly was seen in 11% and 18.4% of all coronary anomalies in similar earlier studies conducted in Turkey and central Europe.⁶

A faulty connection between the coronary artery and another vessel/chamber is known as coronary artery fistulas (CAFs). It is a rare congenital anomaly with a reported incidence of only 0.002% in the general population.³¹ The coronary steal phenomenon occurs due to the diversion of blood from the high resistance myocardial capillary bed into the low resistance fistula.³² Our study had 2 cases of CAFs (2.2% of patients). In the earlier study, it was found out that more than 50% of the times, CAFs involve the RCA.³³ However, in the present study, we found that all CAFs involved the LAD. Moreover, inconsistent with previous reports showing that most of CAFs shunt into the right ventricle and right atrium,³⁴ our study showed one CAF shunted into the left atrium and the other into the right atrium. CAFs can cause myocardial ischemia, syncope, and sudden death. They are treated by surgical ligation, coil occlusion, and covered stent.

In our study, we didn't find other coronary anomalies like, anomalous origin of LMCA arising from the RSV, anomalous origin of LAD from the RSV/RCA, anomalous origin of LCx from the RCA, single coronary artery.

Our study has several limitations. First, the design of the study is retrospective owing to which some important clinical characteristics may not have been recorded. Second, it is a single-centered study. Third, the courses of the anomalous coronary arteries were not defined.

CONCLUSION

The prevalence of coronary anomalies in this study was 1.4%, which is similar to that of previous

studies. Coronary anomalies are hence rare. However, as some types of coronary anomalies are clinically significant and sometimes life threatening, it is important that they are detected in angiographic images. Finally, understanding coronary anomalies can aid physicians in treatment planning.

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CONFLICT OF INTEREST

The author(s) declare that they do not have any conflict of interest with respect to the research, authorship, and/or publication of this article.

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