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Clinical and Angiographic Profile of Coronary Artery Anomalies in Patients Undergoing Coronary Angiography

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Abstract

Introduction: Coronary artery anomalies are relatively rare congenital abnormalities that are often found incidentally during coronary catheterisation done for various indications.Aim of the study was to study clinical and angiographic profile of various coronary anomalies in out institute with respect to demographic profile, frequency of coronary anomalies, atherosclerotic coronary artery disease involvement of anomalous coronaries and associated cardiac co-morbidities. *Methods:* In this study total 162 patients who were reported as having coronary anomaly from 2011 January to 2014 March were evaluated. These patients had underwent coronary angiogram for various indications. All patientsdata including demographic profile, clinical presentation, ECG, 2D echo, routine biochemical investigation reports were analysed and angiograms were reviewed. Various coronary anomalies were classified and relative frequency calculated and cardiac co-morbidities were analyzed. *Results:* Out of 25023 coronary angiograms done during study period, 162(0.65%) patients were found to have coronary anomalies. Among 162 patients, 134(82.7%) were male and 28(17.3%) were female. Average age of presentation was 54.4 years. Most of these patients had conventional risk factors like hypertension (33.9), diabetes mellitus (30%), dyslipidaemia (18.5%), smoking (33.3%) and family history of ischemic heart disease(7.4%). 6.7% patients with coronary anomalies had past history of ischemic heart disease. Among 162 patients, 108(66.7%) presented with acute coronary syndrome, 37(22.8%) with chronic stable angina and in 17(10.5%) patients coronary angiogram was done for other indications. *Conclusion:* Angiographic incidence of coronary anomalies in patients undergoing coronary catheterization in present study is 0.65%. Most of the anomalies were low risk coronary anomalies diagnosed incidentally. Anomalous coronary arteries are frequently involved in atherosclerotic coronary artery disease and have conventional risk factor profile and clinical presentations.

Keywords: Coronary Artery Anomaly; Coronary Angiography.

Introduction

Normal coronary artery anatomy encompasses a wide spectrum. Minor variations, such as number of septal perforator or diagonal branches and size and distribution of individual vessels, ensure that no two anatomic patternsare exactly alike. Coronary artery anomalies representmarked deviations from the normal pattern. Anomalies are present at birth, but relatively few aresymptomatic during childhood. Most anomalies are discoveredas incidental findings during coronary arteriographyor at autopsy. However, some anomalies presentwith symptoms or

potentially serious sequelae that requiresurgical treatment. The clinician should suspect thepresence of a coronary artery anomaly in a young personwho experiences exertional syncope, myocardial infarction, exercise-induced arrhythmias, or cardiac arrest. The angiographer should be able to identify anomaliesin order to perform accurate evaluations and avoidsubsequent errors in management. The cardiac surgeonmust be aware of the abnormal anatomy in order to avoid accidental ligation or transection at the time of surgery [1].

Primary congenital coronary anomalies are detected in about 0.29–1.34% of adult patients undergoing coronary arteriography [1,3,6-14]. Geographic variations in the frequency of different coronary anomalies are well known. There is paucity of Indian data on this aspect.

This study is intended to define the anatomical patterns, frequencyof occurrence, and clinical significance of coronary artery anomalies in patients studied angiographically in our institute.

Methods

Study Design and Protocol

The data were collected retrospectively by analyzing the angiographic data of 25023 consecutive adult patients undergoing coronary angiography between January 2012 and March 2014 at Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bengaluru, India. Coronary angiographies were indicated for stable coronary artery disease in 22.8% of the patients, acute coronary syndromein 66.7% of the patients, valvular heart disease in 6.8 % of the patients, and for other reasons in 3.7% (congenital heart disease, hypertrophic cardiomyopathy, complete heart block etc.) of the patients. The catheterization reports were analyzed, and thosewith anomalous coronary arteries were selected for further assessment. The angiograms were reviewed before finally being classified. In case of any difference of opinion, a consensus wasreached after discussion. Patients with coronary anomalies associated with complex congenital heart disease were not included in this study.

Patients were categorized as having single, double or triple vessel disease when a significant lesion (defined as more than 50% narrowing of intraluminal diameter) was present in one or more coronary arteries or in a major branch. The selective cannulation of aberrant arteries especially origin of RCA from left coronary sinus can be difficult and time-consuming. For thispurpose hockey stick, multipurpose, right and left Amplatz, Judkins right and left catheters were used for angiography.

Eligibility Criteria

Inclusion Criteria

1. Patients who underwent coronary angiogram at SJICR between January 2012 and March 2014 and who were diagnosed as having coronary artery anomalywere included in the study.

Exclusion Criteria

1. Patients with separate origin of the Conus branch
2. Patients with right ventricular branch from the right sinus of Valsalva
3. Patients with coronary anomalies occurring as part of complex congenital heart disease

Results

A total of 162 patients diagnosed as coronary artery anomaly were evaluated in the present study. Among 162 patients, 108 (66.7%) presented with acute coronary syndrome, 37(22.8%) with chronic

Table 1: Clinical presentation

Presentation	Number of Patients	Percentage (%)
Chronic stable angina	37	22.8
Unstable angina	9	5.6
NSTEMI	13	8.0
AWMI	34	20.9
IWMI	23	14.2
IW+RVMI	12	7.4
IW+PWMI	9	5.6
LWMI	8	4.9
RHD	9	5.6
Others	8	4.9
Total	162	100

angina and in 17(10.5%) patients coronary angiogram was done for other indications. Among acute coronary syndrome, 9 (5.6%) patients presented with unstable angina, 13 (8.0%) with NSTEMI and 86 presented with STEMI. Among STEMI, 34 patients presented with AWMI (20.9%), 44 presented with IWMI (27.2%) and 8 patients with lateral wall myocardial infarction (4.9%).

Among 37 patients with chronic angina, 36 had effort angina and one had atypical angina. Valvular heart disease was indication for coronary angiogram in 11 patients with 9 (5.6%) patients of Rheumatic heart disease and 2 patients of Calcific severe aortic stenosis. Other patients included 2 patients of hypertrophic cardiomyopathy, complete heart block in 1 patient, WPW syndrome in 1 patient, atrial septal defect in 1 patient and noncardiac chest pain in 1 patient.

Among 162 patients, most of the patients were in the age group of 41 to 70years. There were 3 (1.85%) patients who were less than 30 years, 17 (10.49%) patients between 31 to 40 years, 43 (26.54%) patients between 41 and 50 years, 50 (30.86%) patients between 51 and 60 years, 40 (24.69%) patients between 61 and 70 years and 9 (5.56%) patients were above 71 years as shown in Table 2. Average age of patients in the present study is 54.4years.

Among 162 patients studied, 134 patients were male and 28 patients were female as shown in table 3.

Among 162 patients, most of the patients had conventional risk factors in the form of hypertension in 55 (33.9%), diabetes mellitus in 49 (30.2%), dyslipidaemia in 30 (18.5%), history of smoking in 54 (33.3%) and family history of CAD in 12 (7.4%) patients as per Table 4. 14 patients had known IHD in the form of previous Percutaeneous Coronary Intervention, Coronary Artery Bypass Grafting or Acute Coronary Syndrome as shown in Table 4.

Table 2: Age distribution of patients studied

Age in Years	No. of Patients	%
<30	3	1.85
31-40	17	10.49
41-50	43	26.54
51-60	50	30.86
61-70	40	24.69
71-80	9	5.56
Total	162	100

Table 3: Gender distribution

Gender	No. of Patients	%
Male	134	82.7
Female	28	17.3
Total	162	100.0

Table 4: CAD risk factor profile in patients with CAA

Risk Factor	No of Patients	Percentage
Hypertension	55	33.9
Diabetes mellitus	49	30.2
Dyslipidaemia	30	18.5
Smoking	54	33.3
Family history of CAD	12	7.4
Known IHD	14	8.6

Table 5: Atherosclerotic coronary artery disease

	Total Number	Percentage
Total number of patients	162	100
Insignificant coronary lesions/normal coronaries	89	54.9
Significant coronary artery disease	73	45.1
Single vessel disease	52	32.1
Double vessel disease	24	14.8
Triple vessel disease	11	6.6
LMCA disease	2	1.2
Anomalous coronary involvement	42	25.9

Out of 162 patients studied, 73 (45.1%) patients had significant coronary artery disease. Among these 73 patients, 42 (25.9%) patients had significant lesions in the anomalous coronary arteries as shown in Table 5.

Among 162 patients with coronary artery anomalies, 98(60.5%) patients had coronary anomalies of origin, 52(32.1%) patients had myocardial bridging, 4 (2.4%) patients had coronary AV fistula, 3 (1.8%) patients had single coronary artery, 3 (1.8%) patients had absent Left main coronary artery, 1 (0.6%) patient had super dominant left anterior desceding artery in which PDA and PLV were arising from large septal branch of Left anterior

Table 6: Distribution of coronary artery anomalies

CAA	No of Cases	Percentage of total CAA	CAD of CAA (%)
RCA from LCS	44	27.2	14(31.8)
RCA fromNCS and High/post origin	20	12.3	8(40)
RCA from LCX	1	0.6	0
RCA from LAD	1	0.6	1
DUAL RCA	1	0.6	0
LCX from RCS	19	11.7	1(5.3)
LCX from RCA	6	3.7	2(33.3)
LAD from RCA	1	0.6	0
LAD from RCS	1	0.6	0
PDA,PLV from S1 of LAD	1	0.6	0
LMCA from NCS	2	1.2	2(100)
LMCA from RCS	2	1.2	0
ALCAPA	1	0.6	0
LMCA absent	3	1.8	0
SINGLE CORONARY	3	1.8	0
myocardial bridging	52	32.1	14(26.9)

Table 7: Anatomic location of myocardial bridging

Location	Number of Patients	Percentage
LAD-proximal	2	3.8
LAD-mid	39	75
LAD-distal	6	11.6
LCX-proximal	1	1.9
LCX-distal	4	7.7
RCA	0	0
Total	52	100

Table 8: Angiographic incidence of coronary anomalies in previous studies

Author/year of publication	Prevalence (%)	Most common coronary anomaly	Imaging modality	Country
Yamanaka et al[1]. 1990	1.30 (1,686 of 126,595)	Absent LMCA with separate origin of LAD and LCX	CAG	USA
Kaku et al[6]. 1996	0.31 (56 of 17,731)	Anomalous origin of RCA from LCS	CAG	Japan
Kardos et al[3]. 1997	1.34 (103 of 7,694)	Absent LMCA with separate origin of LAD and LCX	CAG	Central Europe
Garg et al[7]. 2000	0.95 (39 of 4,100)	Anomalous origin of RCA from LSV/NCS	CAG	India
Yildiz et al[8]. 2010	0.90 (112 of 12,457)	Absent LMCA with separate origin of LAD and LCX	CAG	Turkey
Erol et al[9]. 2011	1.96 (53 of 2,096)	Absent LMCA with separate origin of LAD and LCX/Origin of RCA from LCS	64MDCT	Turkey
Fujimoto et al[10]. 2011	1.52 (89 of 5,869)	Anomalous origin of RCA from LSV	64MDCT	Japan
Sivri et al[11]. 2012	0.74 (95 of 12,814)	LCX arising from RSV or RCA	CAG	Turkey
Sohrabi et al[12]. 2012	1.30 (79 of 6,065)	Absent LMCA with separate origin of LAD and LCX	CAG	Iran
Xu et al[13]. 2012	1.02 (124 of 12,415)	Anomalous origin of RCA from LCS	Dual-source CTCA	China
Yukel et al[14]. 2013	0.29 (48 of 16,573)	Anomalous origin of LCX from RCA/RSV	CAG	Turkey
Present study	0.65(162 of 25023)	Anomalous origin of RCA from LCS	CAG	India

descending artery and 1(0.6%) patient had dual right coronary artery.

The most common coronary anomaly of distribution in present study was intramural coronary artery(myocardial bridging). Total of 52 patients had myocardial bridging anomaly out of 162 patients. 75% of these were present in mid part of Left anterior descending artery, 11.6% in distal LAD, 3.8% proximal LAD, 7.7% in distal LCX, 1.9% in proximal LCX and none in RCA

Discussion

Most of the coronary artery anomalies are asymptomatic.The prevalence of coronary artery anomalies shows a wide variation. They are usually encountered as coincidental findings during coronary angiography or at autopsy. In different studies, the angiographic incidence was reported as 0.29% to 1.34% [1,3,6,7-14] as shown in Table 8.

In our study there was 0.65 %(total of 162 cases) incidence of coronary artery anomalies among 25,023 patients undergoing coronary angiography for various indications.

Recently one more study by Angeliniet al [15] demonstrated high incidence of coronary artery anomalies of 5.64% by collecting magnetic resonance imaging data in a large population of School children. Taking into consideration of minor coronary anomalies which could be detected by MRI may partially responsible for such high incidence. One more explanation is that significant number of coronary anomalies may end in sudden cardiac death in early life thereby reducing the prevalence in elderly.

Coronary artery anomalies could lead to life threatening symptoms, including arrhythmias, syncope, myocardial infarction, or sudden death. In one study, 33% of patients

with anomalous coronary anatomy had a preceding history of angina and or syncope [1]. In our study, among 162 patients, 108(66.7%) presented with acute coronary syndrome, 37(22.8%) with chronic angina and angiographically 42(25.9%) patients had significant lesions in anomalous arteries.

The origin and distribution anomalies of the coronary arteries in our study was 97.5% and coronary artery fistulaewas 2.5%, with somewhat lower incidence of coronary artery fistulas compared to previous studies (95% and 5%,respectively) [1,4,5]. The most common anomaly of origin of coronaries in our study group was origin of RCA from Left Coronary Sinus in 44 (27.2%) patients similar to few earlier studies as can be seen in Table 8. The origin of RCA from noncoronary sinus and/or high posterior origin was second most common seen in 20 (12.3%) patients and origin of LCX from Right Coronary Sinus was third most common among anomalies of origin seen in 19(11.7%) patients. Absent LMCA and separate origin of LAD and LCX was most common anomaly in few earlier studies [1,3,8,9,12] and was seen only in 3 cases in the present study.

The anomalous origin of the LMCA from the right coronary sinus of Valsalva is a rare congenital coronaryanomaly [1,16-18]. This anomaly was found in only two patients in our study with incidence of 1.2% among patients with coronary artery anomalies.

The LMCA, LAD, and RCA may arise from the pulmonary artery in the order of decreasing incidence. Unfortunately, about 90% of patients with these anomalies die during Infancy [19].There was such patient with Anomalous origin of left coronary from pulmonary artery (ALCAPA) in present study who was a 45 year old female with severe left ventricular dysfunction demonstrating exceptional survival.

There were three cases of Single coronary artery in the present study. In all these patients the single coronary artery arising from right sinus of Valsalva and giving rise to all three coronaries. Among these two cases were of Lipton type R-II and one case was of type R-III [37].

The incidence of the LCX arising from the RCA was seen in 6 patients making incidence of 3.7%, isgenerally believed to be of no clinical significance [20]. However 2 of such patients had obstructive coronary disease in our study.

The LMCA or LAD arising from the right coronary sinusof Valsalva and RCA originating from the left coronarysinus of Valsalva deserve clinical attention because these anomalies may be associated with sudden cardiac death inotherwise normal individuals [20-24]. In a study of 12,457 patients by Yildiz A et al [8] 11 such anomalies patients were seen (LMCA fromright sinus of Valsalvain 1 patient, RCA from the left coronary sinus of Valsalva in 10 patients). In present study total of 46 such anomalies were present with LMCA from RCS in 2 patients and RCA from LCS in 44 patients, latter making the most common coronary anomaly of origin.

The anomalous origin of the LMCA from the right coronary sinus of Valsalva can be subclassified into 4 types based on the relationship of the LMCA to the great vessels: septal (beneath the right ventricular

infundibulum), anterior tothe pulmonary trunk, retroaortic, and interarterial (between the pulmonary trunk and the aorta). Roberts et al observed that the interarterial course was found in 60% of patients in whom the leftmain coronaryartery originates from the right coronary sinus [25]. Patientswith this form of LMCA anomaly usually suffer from premature cardiac death [23,26]. Reduction of the coronary blood flow resulting from compression over LMCA from the right coronary sinus of Valsalva or th efirst segment of a single coronary artery suchas the RCA from the left coronary sinus of Valsalva or LAD, coursing interarterially, by the pulmonary trunk or aorta during pressure increase in these vessels upon exercise in addition to acute take off or slit-like orifices of these arteries,produce ischemia resulting in angina, syncope, congestive heart failure, arrhythmias, and sudden death [27-31]. Computed tomographic angiography is a safe and effective noninvasive imaging modality for defining these coronaryanomalies and provides detailed 3-dimensional anatomic information that may be difficult to obtain with invasive angiography [20].

Basically the 3 benign forms of the anomalously originating LMCA from the right coronary sinus of Valsalva donot require specific interventions or cessation of sports activities,while interarterially coursing LMCA may warrant surgical repositioning, especially if associated with objective evidence of ischemia [31]. It is important to differentiate this interarterial coursefrom intraseptal (which may appear similar, but the anomalous vessel passes more inferiorly within themuscular septum) and the posterior course that haverelatively benign courses than the first one [20]. Because of the unusual location and the noncircular coronary orifice of these anomalies, selective catheterization and percutaneous intervention can be technically challenging, particularly with regard to adequate guide catheter support. In cases of anomalous origin of right coronary artery from left coronary sinus, intervention is generally indicated only if symptomatic.

Intramural coronary artery (myocardial bridging) was most common CAA in the present study which was seen in 52 (32.1%among CAA and 0.2% of all patients) patients. Myocardial bridging was seen most commonly in mid LAD(75%) followed by distal LAD, distal LCX, proximal LAD and proximal LCX in decreasing frequency and was not seen in RCA in the present study. These finding were similar to those reported in previous studies [35]. 14 of 52 patients having myocardial bridging had significant obstructive coronary lesions. Endothelial dysfunction is frequent in patients with myocardial bridges. Coronary stenting in these cases associated with high restenosis rate and stent crushing [36].

Coronary artery fistulas (CAF) are rare congenital anomalies. The incidence is around 0.002% in thegeneral population and 0.3% to 0.4% in patients with congenital heart defects [32-33]. The CAF in the present study were seen in 4(0.016%) patients among patients undergoing diagnostic coronary angiography. Among these two were arising from RCA and 1 each from LAD and LCX. Two of these were draining to RA, one to PA and one to PDA. Large CAF can cause important coronary morbidity and mortality leading to angina, syncope, congestive heart failure, myocardial infarction, and sudden death. Surgical ligation is the standard treatment for symptomatic CAF, but is associated with higher morbidity. In the last decade, percutaneous methods have developedas an alternative treatment modality [34].

Study Limitation

Only patients who had undergone coronary angiograms and diagnosed as coronary anomaly were included rather than a randomly selected sample of thewhole population

Conclusion

Coronary artery anomalies are rarely identified during life, often because of insufficient clinical suspicion. Familiarity with coronary artery anomalies may be useful for dealing with diagnosis and treatment of these pathologies especially anomalous origin of coronary artery from opposite sinus of Valsalva, which have clear ischemic potential.

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