



Prevalence Rates of Congenital Coronary Anomalies and Coronary Variations in Adult Indian Population Using Dual-Source Computed Tomography Coronary Angiography: Analysis of Regional Distribution of Coronary Anomalies and the Need for Standardized Reporting Formats

Kartik P. Ganga^{1,*} Aayush Goyal^{2,*} Vineeta Ojha^{1,*} Siddharthan Deepti³ Sanjiv Sharma¹
Sanjeev Kumar¹

¹Department of Cardiovascular Radiology and Endovascular Interventions, All India Institute of Medical Sciences, New Delhi, India

²Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India

³Department of Cardiology, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence Sanjeev Kumar, MD, Department of Cardiovascular Radiology and Endovascular Interventions, All India Institute of Medical Sciences, New Delhi 110029, India (e-mail: sanjeevradio@gmail.com).

Indian J Radiol Imaging 2021;31:138–149.

Abstract

Background Congenital coronary artery anomalies (CCAA) are predominantly discovered as incidental findings on computed tomography coronary angiography (CTCA) of adults. They are rare but significant, considering their importance during endovascular or surgical interventions. This study describes the prevalence of CCAA and coronary variants (CV) in adults as identified by CTCA.

Methods It is a retrospective evaluation of 7,694 CTCAs of adults performed in a tertiary care facility in North India.

Results CCAA and CV were observed in a total of 9.6% of patients. The most common CV was myocardial bridging, observed in 7.1%. Anomalies of origin and course were detected in 2.3% of the patients. The frequency of these anomalies in the right coronary artery, left main, left circumflex artery, and the left anterior descending artery arteries were 1.06, 0.41, 0.03, and 0.38%, respectively. The single coronary pattern was seen in 0.05% and coronary artery fistulas in 0.03%. Scrutiny of data on Indian regional distribution revealed differing definitions and inclusion and exclusion criteria, making comparisons difficult, highlighting the need for uniform definitions as well as the need to adopt a standardized reporting template and format.

Conclusion The prevalence of CCAA and CV is 9.6% in adult Indian patients undergoing CTCA. Prior knowledge of these anatomical finding can prevent a catastrophe during surgery or endovascular interventions. Hence, it is important that clinicians, as well as radiologists, are aware of these entities.

Keywords

- ▶ computed tomography coronary angiography
- ▶ coronary artery anomalies
- ▶ coronary variations

*These authors contributed equally.

DOI <https://doi.org/10.1055/s-0041-1730135>
ISSN 0971-3026

© 2021. Indian Radiological Association

This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (<https://creativecommons.org/licenses/by-nc-nd/4.0/>).

Thieme Medical and Scientific Publishers Private Ltd. A-12, Second Floor, Sector -2, NOIDA -201301, India

Introduction

Congenital coronary artery anomalies (CCAA) and natural coronary variants (CV) are rare. They may be observed in isolation or can be associated with congenital heart diseases,¹ and maybe discovered incidentally on catheter angiography or computed tomography coronary angiography (CTCA) in adults. Sometimes they are diagnosed during endovascular or surgical intervention performed for other causes and rarely during autopsy. Only ~20% of CCAA cause symptoms of angina, syncope, dyspnea, ventricular arrhythmia, heart failure, or acute coronary syndrome.² They may even result in sudden death in young athletes at peak exertions. CCAA or CV vessels may also have increased susceptibility to atherosclerosis.³

Their clinical significance lies in forewarning the operator, either the surgeon or interventionalist, who will be able to negotiate the challenge successfully with the right tools. Catheter angiography in these rare variations, if not detected pre-procedurally, is not only challenging but have an increased fluoroscopic time as well.⁴ They are also of paramount significance in the current scenario where endovascular replacement of aortic valves is becoming more common. A surgeon, on the other hand, may face difficulty during coronary artery bypass grafts (CABG) and may unintentionally ligate the anomalous vessel or there may be inadvertent compression of the coronary artery post valve replacement surgery.⁵

Their overall prevalence rates in India are unclear and variable as are the classifications systems owing to its diversity and differing definitions. As our understanding improves, the CCAA/CV definitions and nomenclatures are revised from time to time, based on which their diagnostic workup, need for intervention, and prognosis are amended.⁶ The distinction between CCAA and CV is debatable and often reported variably. The two parameters that are usually taken into consideration in deciding between labeling an abnormal finding as a CCAA or CV are (1) morphological features (e.g., number of ostia, course, or termination) and (2) frequency of their occurrence. The definitions that are considered appropriate by most authors are (a) if any morphological feature is present in more than 1% of a population, it is deemed as normal; (b) any morphological form that is seen in less than 1% of the population is considered as an anomaly; (c) a normal variant is the occurrence of an unusual morphological feature (e.g., myocardial bridge), but the prevalence is seen in more than 1% of the population.³ It is worth highlighting that CV, because of its prevalence in more than 1% population, may be considered normal in any binary classification system but is not considered so per se because of the current understanding that this morphological finding, although common, is still not a normal feature of a coronary artery.

These definitions are based on the prevalence rates in the population and hence call for large volume data analysis. Currently, there is a scarcity of good-quality large-scale Indian data, particularly derived from cross-sectional modalities that identify more anomalies. Also, there exists no region-wise or modality-wise distribution of these

anomalies in the vast and often heterogeneous Indian population. Our study assessed the prevalence rates of CCAA and CV using CTCA and our results were then compared with prevalence rates from other anatomical and imaging (both cross-sectional and conventional angiography) studies from across India to get a wider perspective of the prevalence rates in India. This would take us one step ahead in forming Indian definitions for CCAA and CV in the adult population and assess if they are in concordance with published world literature. We also seek to emphasize the need for a reporting template for standardizing the CTCA reporting across the country, specifically with regard to CCAAs and CVs in order for better data-driven approaches in assessing the prevalence rates and clinical significance of these entities in the future.

Methods

This study is a retrospective analysis of 7,694 consecutive CTCAs performed in patients aged more than 18 years. It is a single-center study at a tertiary level, cardiac referral service and included the adult CTCAs performed between January 2015 and December 2018. CTCAs performed for the assessment of congenital anomalies were excluded. All CTCAs were acquired using either a dual-source 128 slice (Siemens Somatom Definition, Erlangen, Germany) or a dual-source 384 slice CT (Siemens Somatom Force, Erlangen, Germany). All the scans were electrocardiography gated and performed after intravenous injection of 1 mL/kg of iodinated contrast media using a bolus track method that automatically triggered the scanning when the attenuation at the descending thoracic aorta reached 100 HU. Images were reconstructed using a B26f convolution kernel, a matrix size of 512 × 512 and 0.6 mm slice thickness. CCAA and CV are classified using the system proposed by Angelini, modified to allow omissions where no cases were recorded^{6,7} (► **Table 1**). A normal coronary artery system should have coronary ostia, located in the respective left coronary sinus (LCS) and right coronary sinus (RCS) in the upper-mid section. This gives origin to the left main (LM) trunk and right coronary artery (RCA), respectively. Each proximal segment has a 45 to 90-degree angle in relation to the aortic wall. The coronary vessel should travel from ostia to its termination, the course being subepicardial and extramural. It must give branches to the dependent myocardium within its territory that are right ventricular (RV) free wall, anteroseptal, and left ventricular (LV) free wall. Myocardial bridging, which is defined as part of the coronary artery travelling in an intramural course, was divided as superficial or deep, based on the thickness of the overlying cardiac musculature (using 2 mm as a cutoff). The coronary dominance (CD) pattern that was reported as right:left:codominant (R: L:C), sex distribution, age distribution, presence/absence of disease in the involved segment in all patients with abnormalities were also recorded.

Pertinent Indian studies (both anatomical and imaging)^{5,8-22} dealing with this subject were reviewed, and studies that included more than 50 subjects were included. Due to the variability of CCAA/CV definitions as well as inclusion/exclusion criteria, the overall prevalence rates are

Table 1 Classification and definitions of coronary artery anomalies

	Type	Definitions/Comments
A	Anomalies of course and origin	
1	Absent LM	Separate origin of LAD and LCX
2	Anomalous location of coronary ostium within aortic root or near proper aortic sinus	
	a. High	Ostium > 10 mm from sinotubular junction
	b. Low	Ostium located at lower end of aortic sinus
	c. Commissural	Ostium located within 5 mm of the aortic valve apposition at the aortic annulus
3	Anomalous location of coronary ostium outside normal "coronary" aortic sinuses	AA/LV/RV/PA/aortic arch/other systemic arteries/noncoronary sinus
4	Anomalous location of coronary ostium at improper sinus	
	a. RCA that arises from left anterior sinus	Retrocardiac/retroaortic/between aorta and pulmonary artery/intraseptal/anterior to pulmonary outflow/posteroanterior interventricular groove
	b. LAD that arises from right anterior sinus	Between aorta and pulmonary artery/intraseptal/anterior to pulmonary outflow/posteroanterior interventricular groove
	c. LCX that arises from right anterior sinus	Retroaortic/posterior atrioventricular groove
	d. LCA that arises from right anterior sinus	Retrocardiac/retroaortic/between aorta and pulmonary artery/intraseptal/anterior to pulmonary outflow/posteroanterior interventricular groove
	e. Single coronary artery	Single coronary artery supplying the entire myocardium—single artery with 2 major branches, 1 of which has a retroaortic course, 2 major branches, 1 of which has an interarterial course, 2 major branches, 1 of which has a prepulmonic course, 3 equally dominant major branches
B	Anomalies of intrinsic coronary arterial anatomy	
1	Congenital ostial stenosis or atresia	Ostial dimple in a coronary sinus without a patent arterial lumen with/ without a fibrotic ostial ridge
2	Coronary ostial dimple	Dimple of an undeveloped coronary ostium with two normally arising coronaries
3	Coronary ectasia or aneurysm	Focal saccular or fusiform dilatation of the coronary artery that is 1.5 times the diameter of adjacent normal segment
4	Absent coronary artery	Descriptive
5	Coronary hypoplasia	Congenital underdevelopment of one or more major branches of the coronary arteries with almost pin point lumen with/without markedly decreased length
6	Intramural coronary artery (muscular bridge)	Coronary artery segment that has a tunneled, intramuscular course
7	Subendocardial coronary course	Descriptive
8	Coronary crossing	Crossing of one artery over the other in the epicardium
9	Duplicated RCA/LAD	Descriptive
10	Anomalous origin of branches (PDA, septal branch or conal artery)	Descriptive
c	Anomalies of coronary termination	Draining into RA/RV/CS/SVC/PA/PV/LA/LV/Multiple
D	Anomalous anastomotic vessels	

Abbreviations: AA, ascending aorta; CS, coronary sinus; LA, left atrium; LAD, left anterior descending artery; LCX, left circumflex; LM, left main; LV, left ventricle; PA, pulmonary artery; PV, pulmonary vein; RA, right atrium; RCA, right coronary artery; RV, right ventricle; SVC, superior vena cava.

not comparable for all studies. Hence, the rates for each specific CCAA/CV were calculated and documented when available. Average modality wise and region wise prevalence rates were also calculated (from 16 Indian studies published in the literature.^{5,8-22}

Results

Totally, 7,694 reports were analyzed for the study, which included 4,968 males and 2,726 females. The prevalence of CCAA and CV was 9.6% ($n = 739$), comprising of 466 males

(63%) and 273 (36.9) females, the mean age being 51 years (18–85 years). The prevalence rates, sex distribution, and CD ratios for all the abnormalities are shown in ►Tables 2 and 3. The salient features are as below

A. Anomalies of Origin and Course:

1. *Absent LM*: Absent LM was noted in 32 (0.41%) patients with a preponderant prevalence in males and with 23:30:0 CD ratio.
2. *Anomalous location of coronary ostium within aortic root or near proper aortic sinus of Valsalva*: High position of ostia was seen in 62 patients (0.8%) with a sex distribution of 44:18 (M: F). They were noted involving RCA, LM, and left circumflex (LCX) in 34, 21, and 1 patient, respectively. Both LM and RCA had a high origin in six (0.07%) patients. One case (0.01%) of commissural origin was noted for RCA and LM, respectively. No low ostial origin was seen.
3. *Anomalous location of coronary ostium outside normal "coronary" aortic sinuses*: RCA was noted arising from ascending aorta in two (0.02) patients. There was one patient with left coronary artery arising from the pulmonary artery (PA) (0.01%).
4. *Anomalous location of coronary ostium at improper sinus*: These abnormalities were classified with respect to artery, site of abnormal origin, and course of the abnormal artery (►Table 2). A total of 72 patients (0.93%) had this anomaly in various combinations. Thirty-seven (0.48%) out of the 38 RCAs (0.49%) arising from the LCS had an interarterial course with significant ostial compression in 6 of the 22 cases. This anomaly was seen predominately involving males (M:F-31:7) with a CD distribution of 32:5:1. Two (0.02%) of the four LMs (0.05%) arising from the RCS had an interarterial course that had a significant ostial compression. The other anomalous LMs had a retroaortic course ($n = 1/0.01%$) or anterior course ($n = 1/0.01%$).

LCX was noted arising from the RCS in 28 patients (0.36%) with a sex distribution of 19:9 and a CD ratio of 27:1:0. All of them had a retroaortic course. Other anomalies of origin from the improper sinus or coronary artery are reported in ►Table 2.

5. *Single coronary artery*: Four (0.05%) patients were identified with a single coronary artery, three were arising from the LCS (0.03%), and one from the noncoronary sinus (NCS) (0.01%). None of the single coronary systems were seen arising from the RCS.

B. Anomalies of intrinsic coronary arterial anatomy:

Absent coronary artery was noted only in one patient with an absent LCX (0.01%). Bridging was noted in 550 patients (7.1%) in nearly all major coronary arteries.

Majority of them were superficial and involved the mid LAD (►Table 3). Types 1 and 4 split left anterior descending (LAD) was noted in three (0.03%) patients each (►Figs. 1 and 2).

C. Anomalies of coronary termination:

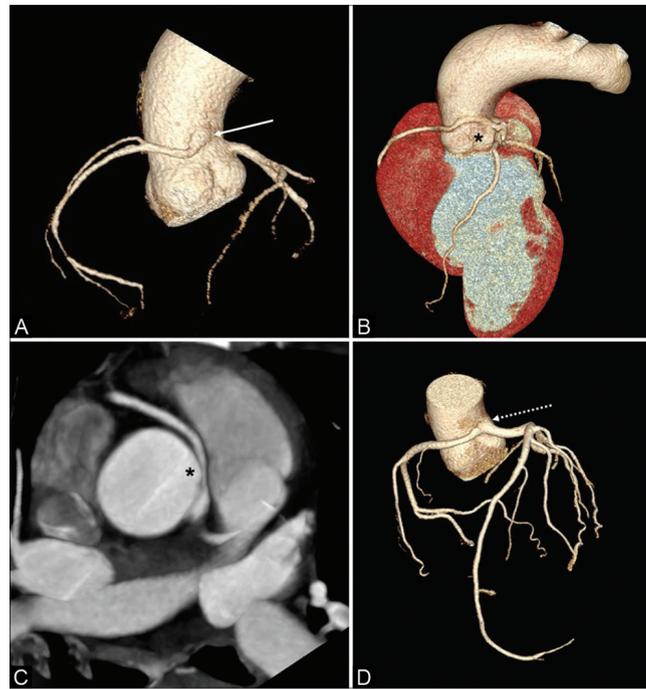


Fig. 1 (A) Volume-rendered computed tomography (CT) image showing high origin of right coronary artery (white arrow); (B) volume-rendered CT image showing right coronary artery arising from left coronary sinus with interarterial course (black asterisk); (C) CT image showing proximal narrowing of a malignant right coronary artery (black asterisk); (D) single coronary artery arising from the left coronary sinus (dashed arrow).

Three coronary artery fistulas (0.03%) were observed (►Fig. 3). Two of them involved the LAD, and one involved the LM. Their terminations were: LAD to RV, LAD to PA, and LM to RV.

Discussion

It is imperative that an epidemiological, anatomical, and morphological descriptive analysis and classification of CCAA and CV must be performed for all populations due to two reasons (a) knowledge about its existence prior to intervention, either endovascular or surgical, prevents inadvertent injury and reduces operator anxiety by allaying surprise, thereby reducing operative time, radiation exposure, and failure rates. It helps in preparing the team by preempting the need for specialized hardware. This is significant in all the cases of catheter angiographies, CABG, valve replacement, and transcatheter aortic valve implantation. (b) Clinical significance of the symptoms in the patients with these anomalies. Most of these, if symptomatic, may manifest in early childhood either stand-alone or in combination with other congenital heart diseases. The interarterial course of an aberrant artery with ostial compression, either dynamic or fixed, may result in cardiac symptoms and even sudden death in adults. Identification of such lesions with long-term follow-up and understanding their prognostic importance may prevent these catastrophic incidents.

Table 2 Prevalence rates of coronary anomalies; coronary dominance represented as right:left:codominant (R:L:C)

	Percentage	Number of cases	Sex ratio (M:F)	Coronary dominance (R:L:C)	Comments
Absent left main trunk	0.41	32	(23:9)	(21:34:0)	
High origin	0.8	62	(44:18)	(55:4:1)	Age—48 y (18–79 y)
RCA	0.41	34	(26:8)	(32:2:0)	Age—49 y (18–73 y)
LM	0.27	21	(14:7)	(18:2:1)	Age 45 y (18–79 y)
LCX	0.01	1	(0:1)	(1:0:0)	Age—73 y
Both RCA and LM	0.07	6	(4:2)	(6:0:0)	Age—49 y (20–69 y)
Commissural	0.02	2	(2:0)	(2:0:0)	
RCA	0.01	1	(1:0)	(1:0:0)	Age—34 y
LM	0.01	1	(1:0)	(1:0:0)	Age—56 y
From ascending aorta	0.02	2	(2:0)	(2:0:0)	
RCA	0.01	1	(1:0)	(1:0:0)	Age—69 y
LM	0.01	1	(1:0)	(1:0:0)	Age—49 y
ALCAPA	0.01	1	(1:0)	(1:0:0)	Age—18 y
RCA that arises from left anterior sinus, with anomalous	0.49	38	(31:7)	(32:5:1)	Age—21 y
Course					
Interarterial	0.48	37			Age—48.19 y (18–70 y) 22 patients with ostial compression—6 significant
Anterior to PA	0.01	1	(1:0)		Age—59 y
RCA from LAD	0.01	1	(0:1)		Age—21 y; prepulmonic course
LCA that arises from right anterior sinus, with anomalous	0.05	4	(1:3)	(4:0:0)	
Course					
Interarterial	0.02	2	(0:2)		Age—47 y (38–57 y); 1 patient with ostial compression—not significant
Anterior to PA	0.01	1	(0:1)		Age—48 y
Retroaortic	0.01	1	(1:0)		Age—52 y
LAD that arises from right anterior sinus, with anomalous	0.02	2	(1:1)	(2:0:0)	Age—74 y
Course					
Interarterial	0.01	1	(1:0)	Age -	26 y; significant ostial compression
Anterior to PA	0.01	1	(0:1)		Age—50 y
LAD from RCA	1	1	(1:0)		Age—78 y; intraseptal course
LCX that arises from right anterior sinus, with anomalous	0.36	28	(19:9)	(27:1:0)	
Course					
Retroaortic	0.36	28			Age—46 y (22–77 y)
LCX from RCA	0.01	1	(0:1)		Age—58 y; retroaortic course
D1 from LCS	0.01	1	(0:1)		Age—47 y
Single coronary artery	0.05	4	(2:2)	(3:1:0)	
From RCS	0.01	1	(0:1)		Age—22 y; RCA with prepulmonic course

(continued)

Table 2 (continued)

	Percentage	Number of cases	Sex ratio (M:F)	Coronary dominance (R:L:C)	Comments
From LCS	0.03	3	(2:1)	Age—42 y (38–49 y); RCA with interarterial course (1 with mild ostial compression)	
Coronary bridging	7.14	550			Age—52.5 y (18–85 y)
Absent coronary artery	0.01	1	(1:0)	(1:0:0)	Age—42 y
Split LAD	0.07	6		(3:0:0)	
Type 1	0.03	3	(2:1)		Age—45 y (32–49 y)
Type 4	0.03	3	(3:0)		Age—43 y (35–53 y)
Anomalies of coronary termination	0.03	3	(3:0)	(3:0:0)	Age—45 y (30–49 y)

Abbreviations: ALCAPA, anomalous LCA from pulmonary artery; D1, diagonal artery; LAD, left anterior descending artery; LCX, left circumflex; LM, left main; PA, pulmonary artery; RCA, right coronary artery.

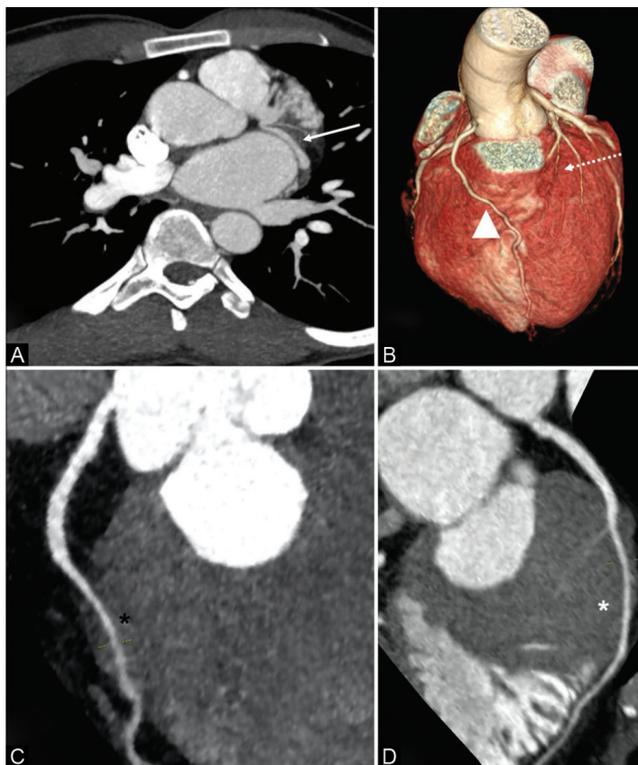


Fig. 2 (A) Computed tomography (CT) image showing circumflex arising from pulmonary artery (white arrow); (B) volume-rendered CT image showing type 4 dual left anterior descending artery, one arising from right coronary sinus (arrowhead) and the other smaller artery arising from left main coronary artery (dashed arrow); (C) curved multiplanar CT image showing short segment bridging of right coronary artery (black asterisk); (D) curved multiplanar CT image showing long segment bridging of mid left coronary artery (black asterisk).

The overall prevalence rates from this adult North Indian population of CCAA/CV were 9.6%. Considering myocardial bridging as a CV, which had a prevalence rate of 7.1%, the other CCAAs had a prevalence of 2.45%. The results are consistent with other Indian studies that report a rate between 1 and 10% (→ Table 4). The high variability could be explained due to exclusion or inclusion of CVs in the different studies and the imaging modality used. Not surprisingly, on average,

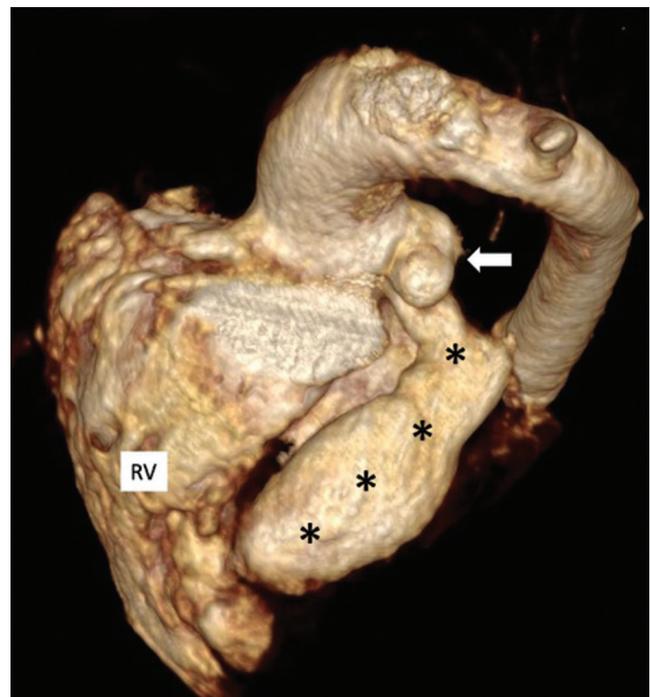


Fig. 3 A volume-rendered computed tomography image showing dilated left main coronary artery arising from left coronary sinus (block arrow) and terminating into the right ventricle (RV), suggestive of coronary artery fistula (black asterisks).

most of the anomalies and CVs were more commonly identified by CTCA as compared with catheter angiography in the studies reviewed. The variability in the prevalence rates between cross-sectional studies and catheter angiography studies can largely be attributed to the superior spatial resolution and reconstructive capabilities of CTCA.

Anomalies of Origin and Course

Absence of LM trunk was observed in 32 patients with a prevalence of 0.48%, similar to the previous Indian studies.^{5,8-22} Considered to be a CV, with insignificant clinical importance, it would, however, be wise to know this finding before CABG. It also bears profound implication to any

Table 3 Myocardial bridging involving the various coronary arteries

	<i>n</i>	Proximal:mid:distal	Superficial:deep	Short:long	Disease in segment (nonsignificant:significant)
LAD	503	16:361:155	492:13	436:34	21:3
LCX	8	6:0:4	8:0	5:3	0:0
RCA	22	1:20:1	22:0	21:1	3:0
RI	4	4:0:0	4:0	3:1	0:0
OM	6	4:0:2	6:0	6:0	0:0
Diagonal	5	4:0:1	5:0	3:1	1:1
PDA	2	1:0:1	2:0	1:1	0:0

Abbreviations: LAD, left anterior descending artery; LCX, left circumflex; OM, obtuse marginal; PDA, posterior descending artery; RCA, right coronary artery; RI, ramus intermedius.

surgery involving the aortic root where either the coronary button is proposed to be harvested or antegrade cardioplegia delivery into the ostium is contemplated. Inadvertent injury, an improper obstructive lie of a reimplanted button or failure to give cardioplegia, may result in catastrophic outcomes.

The same holds true for all the anomalies of coronary origin. Anomalous ostia may either be located within the same original sinus or may be located outside the region of the respective normal coronary sinuses. We observed one commissural origin for the RCA and the LM, respectively, while a high origin was observed in 62 patients amounting to nearly 0.8% of prevalence. RCA was involved in this displacement almost two out of three times, with only one high origin of the LCX noted. Very rarely, improper aortic clamping during surgeries below or at a coronary artery arising from higher than expected location may result in failed cardioplegia as well as injury to the coronary artery. The higher origin may also lead to perfusion disturbances *de novo* due to the absence of the facilitatory effect of the coronary sinus on the diastolic coronary runoff. We did not observe any low coronary ostia, which are exceedingly rare, 0.02% according to the Indian literature. It was less commonly identified in south Indian populations, and its prevalence rates were similar in both CTCA as well as catheter angiography studies (►Table 4).

When the coronary ostia are located outside the sinuses that normally give origin to the coronaries, it may take its origin from various locations including NCS, ascending aorta, arch and its branches, LV, RV, other systemic arteries, and from the PA. We observed one case each of the RCA and LM arising from the ascending aorta and one case of LM arising from the PA.

When a coronary originates from a sinus other than its eponymous sinus, one of the arteries will have an abnormal course. Depending on the artery involved, the abnormal course may be retroaortic, between aorta and PA (interarterial), transseptal, anterior to pulmonary outflow (prepulmonic), or in the posterior atrioventricular groove. We observed an overall prevalence of anomalous location of the coronary ostia at improper sinus in 72 patients (0.93%). The two most common variants were RCA that originated from the LCS and LCX that originated from the RCS, with a prevalence of 0.49 and 0.36%, respectively. It is consistent

with the average prevalence rates of anomalous RCA found in 0.5 and 0.3% identified by CTCA and catheter angiography in Indian literature, with no definite preponderant regional distribution (►Table 4). In the subsegmental analysis of the RCA that arises from LCS, out of the 37 patients with interarterial course, 22 patients had ostial compression, of which six were significant. Among the LM from RCS group with interarterial course, one quarter had ostial compression. We also observed significant ostial compression in the one patient where LAD was arising from the RCS with an interarterial course. Anomalous origin of LCA was more commonly identified using CTCA as opposed to catheter angiography in most Indian studies with very limited region wise data available. The retroaortic course of the LCX, albeit benign, is important for the surgeon, as there is a potential for it to be injured during aortic valve surgery.²³ It is identified more commonly in the south Indian population as opposed to the rest of the country (~0.36 vs. 0.2%) with a tendency to be identified more commonly on CTCA (►Table 4).

Single coronary artery was observed in four patients with three arising from the LCS and one from NCS. Our prevalence of 0.06% is slightly higher than that of internationally reported 0.0024 to 0.044% prevalence rate but consistent with prevalence rates from other Indian populations. Even in this rare subset, the one patient who had interarterial RCA demonstrated ostial compression. As suggested before, the interarterial course may potentially cause sudden death, making it more pertinent in cases of single coronary artery.²⁴ It is more commonly identified in Indian studies using CTCA as compared with catheter angiography with slightly higher average prevalence rates noted in the north Indian population.

We identified only one abnormal origin of the LM from PA system reinforcing their fatal course with patients not surviving till adulthood. This is largely concordant with other studies involving adult populations with no definite modality wise or region wise variation in distribution.

Anomalies of Intrinsic Coronary Arterial System

Myocardial bridging is the presence of a segment of coronary artery lying completely within the myocardium. It is fairly common and is therefore classified as a CV rather than a CCAA.

Table 4 Analysis of regional and modality wise prevalence rates from various regions of India

	Current study	Yadav et al ¹⁸	Sirasapalli et al ¹³	Majumdar et al ¹⁷	Raval et al. ¹⁹	Lingaraju et al. ²⁰	Nawale et al ²¹	Average prevalence rate—CA ^a	Average prevalence rate—CTCA ^a	North India average prevalence rate ^a	South India average prevalence rate ^a	West India average prevalence rate ^a
Year of publication	2020	2015	2018	2016	2016	2016	2018	-	-	-	-	-
Modality	CT, (384 second DS)	CT (128 second SS)	CT (64 second SS)	CA	CA/CT/ECHO	CA	CA	-	-	-	-	-
Age	Adults	All	(10-77)	Adults	All	Adults	Adults	-	-	-	-	-
Exclusions	Ectasia, split RCA, conal artery origins, intracavitary coronaries					High ostia absent LM, bridging, ectasia	Bridging excluded					
Population	North Indian	North	South	South	West	West	West	-	-	-	-	-
Study population	7,694	1,000	8,021	2,3311	17245	8,500	4,481	-	-	-	-	-
CAM	-	3.1	10.09	0.93	1.49	1.27	1.91	-	-	3.20	3.84	1.56
Absent LM	0.41	-	0.18	0.265	0.319	0.42	0.45	0.42	0.90	1.53	0.35	0.40
High origin	0.8	0.2	0.07	-	0.18	-	0.332	0.20	0.11	0.20	0.06	0.26
LM/RCA from Aorta	0.02	0/0.2	-	-	0.017	-	0.02/0.33	-	-	-	-	-
Commissural/low	0.01/0	-	-	-	0/0.023	-	-	-	-	-	-	-
RCA from LCS	0.49	0.8	0.5	0.238	-	0.4	0.45	0.37	0.51	0.42	0.44	0.43
Interarterial	0.48	0.8	-	-	0.151	-	0.45	-	-	0.51	0.39	0.30
Anterior	0.01	-	-	-	0.023	-	0	-	-	-	-	-
LCA from RCS	0.05	-	0.14	0.013	-	0.07	-	0.04	0.27	0.78	0.14	0.07
Interarterial	0.01	-	-	-	0	0.02	-	-	-	-	0.13	0.02
Anterior	0.02	-	-	-	0.006	0.01	-	-	-	-	-	-
Retroaortic	0.01	-	-	-	0.006	0	-	-	-	-	-	-
LAD from RCS	0.02	-	0.01	-	-	0.02	-	0.02	0.09	-	-	-
LCX from RCS	0.36	-	0.19	0.107	-	0.25	0.33	0.24	0.41	0.27	0.37	0.29
Retroaortic	0.36	-	-	-	0.226	0.25	0.33	-	-	-	-	-
LCX from RCA	0.01	-	-	-	-	-	-	-	0.06	-	-	-
ALCAPA	0.01	0.1	-	-	0.029	-	-	0.02	0.25	0.06	0.40	0.03

(continued)

Table 4 (continued)

	Current study	Yadav et al ¹⁸	Sirasapalli et al ¹³	Majumdar et al ¹⁷	Raval et al ¹⁹	Lingaraju et al ²⁰	Nawale et al ²¹	Average prevalence rate—CA ^a	Average prevalence rate—CTCA ^a	North India average prevalence rate ^a	South India average prevalence rate ^a	West India average prevalence rate ^a
Single coronary artery	0.05	0.2	0.12	0.021	0.058	0.02	0.02	0.04	0.13	0.16	0.07	0.03
Absent coronary artery	0.01	–	–	0.008	–	–	–	0.01	0.19	0.19	0.14	–
Split LAD	0.07	0.2	0.06	0.055	0.006	–	0.02	0.25	0.22	0.42	0.06	0.01
Split RCA	–	0.6	–	0.124	0.046	–	–	0.34	0.60	0.72	0.09	0.05
Fistula	0.03	0.2	0.03	–	0.2	0.02	0.11	0.09	0.12	0.12	0.05	0.11
Fistula to RV	0.02	0.1	0.012	–	0.087	0	–	–	0.06	0.10	0.01	0.09
Fistula to RA	–	–	0.012	–	0.075	0	–	0.04	0.01	0.04	0.01	0.08
Fistula to others	0.01	0.1 (pa)	0.012	–	0.041	0.02 (PA)	0.11	0.06	0.01	0.01	0.01	0.08
Anomalous Anastomatic vessels	–	0.1	–	–	0.017	–	–	–	0.10	0.10	–	0.02
Bridging	7.1	–	9.05	–	0.093	–	–	0.13	10.68	19.40	4.26	0.09

Abbreviations: AA, ascending aorta; ALCAPA, anomalous LCA from pulmonary artery; CA, catheter angiography; CS, coronary sinus; CTCA, CT coronary angiography; LA, left atrium; LAD, left anterior descending artery; LCX, left circumflex; LM, left main; LV, left ventricle; PA, pulmonary artery; RCA, right coronary artery; RV, right ventricle; SVC, superior vena cava.

^aThe composite prevalence of coronary anomalies has been calculated from all the Indian studies published in the literature.^{5,8,22} However, only the six largest studies have been represented in the table, other than the current study.

The prevalence widely varies between 0.5 and 16% on catheter angiography and 40 to 80% during autopsy.²⁵ Predictably, CTCA has detected a higher prevalence of 21.3% as compared with catheter angiography.²⁶ We report a prevalence of 7.1% among our patients that is consistent with other Indian studies that used CTCA. Currently, CTCA is able to accurately diagnose bridging, with even the rare intracavitary coronary segment being consistently identified. Although considered a benign entity, it is postulated that sometimes it could be the functionally significant due to systolic compression by overlying musculofibrous tissue resulting in ischemic symptoms.²⁷ This may be associated with the depth or length of bridging, but no definitive evidence is available in the literature. The bridging may be subdivided with respect to the depth and segment of the artery involved. We arbitrarily divided the depth and length as follows (<2 mm—superficial; >2 mm—deep; <5 mm—short; >5 mm—long). The most common vessel that is involved according to the literature²⁸ is the mid-LAD that is coherent with our findings where 503 out of 550 cases of bridging involved the LAD, with >90% involving a short, superficial segment. The presence of bridging is thought to be protective from atherosclerosis. Still, we identified patients with plaques within the bridging segment, with 0.7% being significant, suggesting that bridging is not always a protective factor. We identified bridging in all major coronary vessels (→ **Table 3**).

We observed one patient of absent coronary artery (0.01) and six patients with a split LAD (0.07%). Split LAD (also called dual LADs) variants consist of two branches supplying the LAD territory instead of one single LAD. Split LADs are mainly of four types. In types 1 to 3, the LAD divides into two branches—a short branch that ends in the proximal anterior interventricular groove and a long one that traverses either on the left (type 1) or right (type 2) of the anterior interventricular groove or intramyocardially (type 3) to finally emerge into the anterior interventricular groove. In type 4, the shorter LAD arises from the LM coronary artery, and the longer LAD arises from the RCA and follows prepulmonic, interarterial, or septal course to reach the anterior interventricular groove.²⁹ Multiple variations of these basic types have also been described in the literature. Both anomalies are more commonly identified on CTCA studies with most regions showing similar prevalence rates.

Anomalies of Coronary Termination

Coronary artery fistula had a prevalence of 0.03% in our study, comparable to rates from other Indian studies ranging from 0.06 to 0.2% with most of the regions reporting similar incidence rates. They can involve either of the main coronaries or their branches and when communicating with the right-sided chambers, they may result in insufficient perfusion due to steal. These fistulas draining into the right heart are diagnosed early in life owing to their pathophysiological manifestation and hence not commonly identified in adults. All three of the fistulas in this study involved the left coronary artery, two in LAD and one in LM, with termination of LAD to RV, LAD to PA, and LM to RV.

Importance of CTCA as a Diagnostic Modality and the Need for Standardized Definitions and Reporting Templates

Incidental discovery during a CTCA is the most common mode of identification of CCAA and CV. Often difficulty in catheter angiography prompts an experienced angiographer to look for these anomalies. Their numbers are increasing because of the increasing number of these investigations, improved awareness among specialists, excellent imaging modalities, and reconstruction softwares. CTCA is a sensitive tool for diagnosing CCAA and CV, but with much less radiation as compared with catheter angiography, with the latest CTCAs being performed with submillisievert exposures. It is noninvasive, easier, cheaper, quicker, more readily available and less operator-dependent. CTCA in most studies is able to identify more lesions than conventional angiography. Not only is this due to the higher spatial resolution of CTCA but also due to many of these anomalies being incidentally picked up in asymptomatic patients, reinforcing the notion that many of these anomalies and variations are indeed benign. However, only careful identification and clinical follow up can confirm or refute anecdotal as well as expert opinions both for the “hemodynamically significant” anomalies and “harmless” anomalies such as bridging with data and evidence-driven clinical strategies.

This sharply brings into focus the need for both standardized definitions for all anomalies and a reporting template that can be used for survey purposes as well as archival/follow-up in a national registry. Standardized reporting templates would also be beneficial to the referring cardiologist or surgeon especially in cases of the “hemodynamically significant” anomalies with specific diagrammatic representation of the anomaly in the form of a “surgeons view” for better appreciation of the anatomical positioning prior to the planned surgeries. This would facilitate the surgery with decreased coronary injury rate and procedure time, with a more confident and determined preplanned approach. Apart from making the use of recommended CADRADS (Coronary Artery Disease- Reporting and Data System) template³⁰ for routine CTCAs mandatory, we recommend using a diagrammatic representation of any anomaly using the template as suggested by the Mery et al.^{31,32} CADRADS can convey the desired CTCA report together with the relevant clinical information and further treatment/imaging strategy to the referring clinician with the diagram clearly representing the information that the surgeon needs from his operating perspective. Although the template by Mery et al is targeted at depicting origin anomalies, it is for these anomalies that surgeries are routinely contemplated. Another reason to make this shift to a nationally agreed standardized template is the easy incorporation of all the data into a registry or electronic data capture tools and softwares. This would enable for wider accumulation of data from multiple regions and institutes to make a true assessment of prevalence rates, clinical relevance, and for formulation of follow-up and treatment strategies. A proposed template, along with an explanation, is given as → **Supplementary File 1** (available online only).

Limitations

Since a single-center data may not be truly representative of the general population, we sought to compile data from other large studies in India. Furthermore, CTCA or catheter angiography is performed in specific groups of people for specific indications who visit the hospital for some cardiac complaint, thus introducing a referral bias. Clinicoradiological correlation would have been ideal but was not the objective of this study. Majority of the studies referenced have similar limitations, and zonal or modality-based comparisons are difficult due to differing definitions, inclusion/exclusion criteria, expertise, and other local factors such as regional variations in imaging referrals, cost considerations, and patient awareness/affordability. This is why standardization may be the key area where efforts must be focused.

Conclusion

The prevalence and pattern of CCAA and CV detected using dual-source coronary CTCA are described from a sizeable Indian population. Considering the size and vast ethnicity of the Indian diaspora, regional variations in prevalence rates are expected, but lack of a standardized reporting format hampers the comparability of various studies. This brings into focus the need for a homogenized alpha numeric and pictorial system for reporting of all CTCAs with the aim to assess the epidemiological data for the CCAA and CVs. The magnitude of application and clinical merit of this system may be tested on historical data, revised, and investigated in prospective registries. The in-practice utility must be verified with respect to ease in reporting, benefit to the interventionist/surgeon with the information conveyed, and advantages in data maintenance and sharing. Consequently, CCAA and CV, which often are underreported, may gain their righteous attention.

Financial Support and Sponsorship

Nil.

Conflicts of Interest

There are no conflicts of interest.

References

- 1 Tuzcu EM, Moodie DS, Chambers JL, Keyser P, Hobbs RE. Congenital heart diseases associated with coronary artery anomalies. *Cleve Clin J Med* 1990;57(2):147-152
- 2 Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990;21(1):28-40
- 3 Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. *Circulation* 2002;105(20):2449-2454
- 4 Rigatelli G, Cardaioli P. Endovascular therapy for congenital coronary artery anomalies in adults. *J Cardiovasc Med (Hagerstown)* 2008;9(2):113-121
- 5 Garg N, Tewari S, Kapoor A, Gupta DK, Sinha N. Primary congenital anomalies of the coronary arteries: a coronary: arteriographic study. *Int J Cardiol* 2000;74(1):39-46
- 6 Angelini P. Coronary artery anomalies: an entity in search of an identity. *Circulation* 2007;115(10):1296-1305
- 7 Sundaram B, Kreml R, Patel S. Imaging of coronary artery anomalies. *Radiol Clin North Am* 2010;48(4):711-727
- 8 Yadav A, Buxi T, Sudarshan H, Jayamma SS, Rawat KS, Ghuman SS. Clinico-radiological correlation of coronary artery anomalies in Indian subcontinent. *Astrocyte* 2015;1:277-287
- 9 Rao A, Pimpalwar Y, Yadu N, Yadav RK. A study of coronary artery variants and anomalies observed at a tertiary care armed forces hospital using 64-slice MDCT. *Indian Heart J* 2017;69(1):81-86
- 10 Sinha P, Saxena S, Jethani SL, Khare S, Jain S, Mehrotra N. Major primary congenital coronary artery anomalies: an angiographic study. *J Anat Soc India* 2012;61:172-176
- 11 Sidhu NS, Wander GS, Monga A, Kaur A. Incidence, characteristics and atherosclerotic involvement of coronary artery anomalies in adult population undergoing catheter coronary angiography. *Cardiol Res* 2019;10(6):358-368
- 12 Diwan Y, Diwan D, Chauhan RS, Negi PC. Coronary artery anomalies in North Indian population: a conventional coronary angiographic study. *Natl J Clin Anat* 2017;06:250-257
- 13 Sirasapalli CN, Christopher J, Ravilla V. Prevalence and spectrum of coronary artery anomalies in 8021 patients: a single center study in South India. *Indian Heart J* 2018;70(6):852-856
- 14 Sakthivel MK, Mathew RP, Wakade AD, et al. 128-Slice MDCT angiographic evaluation of coronary artery anomalies in the South Asian (Indian) population – a first experience. *J Indian Coll Cardiol* 2017;7:116-122
- 15 Prasad S, Vijayakumar DH. Detection of coronary artery anomalies using 64 slice MDCT angiography. *Int J Contemp Med Surg Radiol* 2017;2:75-79
- 16 Aiyappan SK, Vadanika V, Karpagam B, Vinayagam S, Saravanan KC. Role of MDCT-Coronary angiography in the evaluation of coronary artery anomalies and incidence of coronary artery anomalies in a subset of South Indian population. *J Evol Med Dent Sci* 2015;4:15561-15565
- 17 Majumdar S, Krishnan H, Kumar A. TCT-488 congenital coronary anomalies of origin and distribution in adult patients referred for coronary angiography. *J Am Coll Cardiol* 2020;68(18, Suppl):B195-B196
- 18 Sivakumar GS, Selvarani G, Kumar SS, Ramesh R, Hemanath TR, Manikandan D. Incidence of coronary artery anomalies among patients undergoing coronary angiography and its relevance to appropriate choice of coronary catheter selection—a tertiary care center study. *Int J Sci Stud* 2017;5:228-231
- 19 Raval A, Jadhav N, Prajapati J, et al. Distribution of coronary artery anomalies and their evaluation with different imaging modalities. *Int J Med Res Rev* 2016;4:1807-1819
- 20 Lingaraju S, Maurya RK, Sanghvi S. A study of incidence and pattern of coronary artery anomalies in Western Rajasthan, India. *Int J Res Med Sci* 2017;4:3388-3393
- 21 Nawale JM, Chaurasia AS, Nalawade DD, Choudalwar P, Borikar N, Tiwari D. Study of clinical profile, incidence, pattern, and atherosclerotic involvement of congenital coronary artery anomalies in adults undergoing coronary angiography: a study from a tertiary care institute in western part of India. *Heart India* 2018;6:133-140
- 22 Srinivasan KG, Gaikwad A, Kannan BRJ, Ritesh K, Ushanandini KP. Congenital coronary artery anomalies: diagnosis with 64 slice multidetector row computed tomography coronary angiography: a single-centre study. *J Med Imaging Radiat Oncol* 2008;52(2):148-154
- 23 Schmitt R, Froehner S, Brunn J, et al. Congenital anomalies of the coronary arteries: imaging with contrast-enhanced, multidetector computed tomography. *Eur Radiol* 2005;15(6):1110-1121
- 24 Kim SY, Seo JB, Do KH, et al. Coronary artery anomalies: classification and ECG-gated multi-detector row CT findings with angiographic correlation. *Radiographics* 2006;26(2):317-333, discussion 333-334

- 25 Shi H, Aschoff AJ, Brambs HJ, Hoffmann MHK. Multislice CT imaging of anomalous coronary arteries. *Eur Radiol* 2004; 14(12):2172–2181
- 26 Arjmand Shabestari A. Coronary artery calcium score: a review. *Iran Red Crescent Med J* 2013;15(12):e16616
- 27 Alegria JR, Herrmann J, Holmes DR Jr, Lerman A, Rihal CS. Myocardial bridging. *Eur Heart J* 2005;26(12):1159–1168
- 28 Ko SM, Choi JS, Nam CW, Hur SH. Incidence and clinical significance of myocardial bridging with ECG-gated 16-row MDCT coronary angiography. *Int J Cardiovasc Imaging* 2008;24(4):445–452
- 29 Spindola-Franco H, Grose R, Solomon N. Dual left anterior descending coronary artery: angiographic description of important variants and surgical implications. *Am Heart J* 1983;105(3):445–455
- 30 Cury RC, Abbara S, Achenbach S, et al. Endorsed by the American College of Cardiology. CAD-RADS™: Coronary artery disease - reporting and data system: an expert consensus document of the Society of Cardiovascular Computed Tomography (SCCT), the American College of Radiology (ACR) and the North American Society for Cardiovascular Imaging (NASCI) *J Am Coll Radiol* 2016;13(12 Pt A) :1458–1466.e9
- 31 Mery CM, Lawrence SM, Krishnamurthy R, et al. Anomalous aortic origin of a coronary artery: toward a standardized approach. *Semin Thorac Cardiovasc Surg* 2014;26(2):110–122
- 32 Molossi S, Martínez-Bravo LE, Mery CM. Anomalous aortic origin of a coronary artery. *Methodist DeBakey Cardiovasc J* 2019;15(2):111–121