ORIGINAL ARTICLE

Significance of Coronary Artery Anomalies and Variants Found on Coronary Computed Tomography Angiography

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ABSTRACT
Objective: We sought to assess the significance of coronary artery anatomical variants and anomalies found in patients referred for coronary computed tomographic angiography (CTA).
Methods: Imaging was done on a 128-detector computed tomography machine followed by three-dimensional reconstruction. Pre- and post-contrast angiographic images of the coronary arteries of 108 patients from January 2013 to December 2016 were retrospectively analysed for coronary artery anomalies and variants.
Results: A total of 74 anomalies and variants were noted in 58 of 108 patients. Among variants, co-dominance, left dominant circulation, ramus intermedius, early origin of posterior descending artery (PDA), and double PDA were noted. Among anomalies, right coronary artery (RCA) origin from the ascending aorta and left coronary sinus, left circumflex artery (LCX) arising from the RCA, malignant course of RCA, retrograde aortic course of LCX, myocardial bridging, and coronary hypoplasia were noted.
Conclusion: Prior assessment for coronary anomalies and variants can guide the interventionalist and surgeon before coronary angioplasty and coronary artery bypass grafting, thereby reducing procedural complications.

Key Words: Aorta; Coronary artery disease; Coronary sinus; Heart defects, congenital; Myocardial bridging; Tomography, X-ray computed

中文摘要
冠狀動脈斷層掃描血管造影術中發現的冠狀動脈異常和變異的意義
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目的：評估在接受冠狀動脈斷層掃描血管造影術中發現的冠狀動脈異常和變異的意義。

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INTRODUCTION
Coronary artery anomalies and variants are in the differential diagnosis of patients with suspected coronary artery disease. A meticulous evaluation of the coronary tree to assess the arteries’ origins and course by electrocardiogram (ECG)-gated coronary computed tomography angiography (CTA) allows accurate analysis with the added advantages of being non-invasive and quick to acquire, with exquisite delineation of cardiac and vascular anatomy. Appropriate depiction of incidentally discovered coronary artery anomalies and variants in individuals with coronary artery disease symptoms is important to prevent unexpected adverse cardiac events.

METHODS
This study was approved by the institutional review board and ethics committee of our institution. This study was conducted at our hospital. This is a retrospective study of the CTA images of patients referred to our hospital for coronary computed tomography (CT) between January 2013 and December 2016. Exclusion criteria were a serum creatinine >1.5 mg/dL, any absolute contraindications to CT or intravenous contrast agents, and suboptimal images.

Procedure
The patients took ivabradine 5 mg BID for 2 days before the scheduled CTA to control and regularise the heart rate for better ECG gating and undistorted images. In selected patients, a slow intravenous injection of metoprolol was given before scanning to maintain the heart rate <70 bpm with minimum beat-to-beat variation.

The CTAs were performed with a 128-detector scanner (GE 660 Optima, General Electric Company, Tokyo, Japan). The technical parameters were: tube current = 400 mAs, tube voltage = 120 kv, tube rotation time = 400 ms, and section thickness = 0.625 mm. Post-image acquisition reformatting and calcium score calculation were performed at a GE 660 ADWOPTIMA workstation.

Frontal and lateral topograms of the chest were acquired to plan the study. Initially, unenhanced CT images were acquired for calcium scoring from the level of the carina to 2.5 cm below the diaphragm with prospective ECG gating. After administration of iohexol (Magnapaque; Magnus Health Management Pvt. Ltd., Mumbai, India) containing 350 mg iodine per mL, 1.5 mL/kg of body weight at a rate of 5.5 mL/s with a double-barrel pressure injector, enhanced coronary angiographic images were acquired with retrospective ECG gating, using the bolus tracking technique.

The images were meticulously analysed with semi-automated commercial software (SmartScore 4.0; GE Healthcare, Chicago [IL], United States) which allows the observer to individually select the calcium foci and label the affected artery, after which the total Agatston calcium score is automatically generated.

Five phases during diastole (50%, 55%, 60%, 65%, and 70%) underwent angiographic analysis after formatting including multiplanar reconstruction, curved multiplanar reconstruction, and three-dimensional volume rendering.

RESULTS
In total, CTA images of 108 patients were identified. We classified the anomalies based on the widely accepted Angelini classification.1 A total of 74 anomalies and variants were identified in 58 of 108 patients; the distribution is shown in Tables 1 and 2. Among origin anomalies (Table 1), a high take-off of the right coronary artery (RCA) from the ascending aorta (n = 2; Figure 1), an RCA originating from the left coronary sinus
Coronary Artery Anomalies on CTA

(n = 1; Figures 2 and 3), and a left circumflex artery (LCX) arising from the RCA (n = 2; Figure 4) were noted.

The course anomalies (Table 1) included malignant course of the RCA between the aorta and the pulmonary trunk (n = 2; Figures 2 and 3), a retroaortic course of

Table 1. Coronary artery anomalies in 108 patients.

<table>
<thead>
<tr>
<th>Origin</th>
<th>No. (%) of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>High take-off RCA</td>
<td>2 (1.9%)</td>
</tr>
<tr>
<td>RCA from LCS</td>
<td>1 (0.9%)</td>
</tr>
<tr>
<td>LCX from RCA</td>
<td>2 (1.9%)</td>
</tr>
<tr>
<td>Course</td>
<td></td>
</tr>
<tr>
<td>Interarterial course of RCA</td>
<td>2 (1.9%)</td>
</tr>
<tr>
<td>Retroaortic course of LCX</td>
<td>2 (1.9%)</td>
</tr>
<tr>
<td>Myocardial bridging</td>
<td>20 (18.5%)</td>
</tr>
<tr>
<td>Coronary hypoplasia</td>
<td>1 (0.9%)</td>
</tr>
<tr>
<td>Total</td>
<td>30 (27.8%)</td>
</tr>
</tbody>
</table>

Abbreviations: LCS = left coronary sinus; LCX = left circumflex artery; RCA = right coronary artery.

Table 2. Coronary artery variants reported in 108 patients.

| Left dominant coronary circulation   | 10 (9.3%)           |
| Co-dominant coronary circulation     | 11 (10.2%)          |
| Double posterior descending artery   | 3 (2.8%)            |
| Early origin of posterior descending artery | 5 (4.6%)        |
| Ramus intermedius                    | 15 (13.9%)          |
| Total                                | 44 (40.7%)          |

Figure 1. Three-dimensional tree virtual reality image showing high take-off right coronary artery (RCA) from ascending aorta (AO; black arrow); sinotubular junction (four-point star), right coronary sinus (RCS), left coronary sinus (LCS), and left coronary artery (LCA).

Figure 2. Multiplanar reconstruction image showing anomalous origin of right coronary artery from left coronary sinus (arrow) and malignant course of right coronary artery between aorta (AO) and pulmonary artery (PA) and right atrium (RA).

Figure 3. Three-dimensional virtual reality image showing anomalous origin of right coronary artery from left coronary sinus (black arrow) aorta (AO), left coronary sinus (star) and malignant course of right coronary artery (RCA) between AO and pulmonary artery (PA; white arrow).
the LCX (n = 2; Figure 4), and myocardial bridging (n = 20) affecting the left anterior descending artery (LAD; Figure 5) and RCA (Figure 6).

One case of coronary hypoplasia was identified (Figures 7 to 10). The left main coronary artery (LMCA) was reduced in its calibre (1.1 mm; normal range, 4.5±0.5 mm in men, 4.0±0.5 mm in women) along its entire length (1.5 cm) before it trifurcated into the LAD, a ramus intermedius artery, and the LCX. The conal artery measured 3 mm in calibre and coursed right to left anterior to the pulmonary conus to anastomose with the LAD distal to the D1 branch forming a Vieussens’ ring (Figures 8 to 10). During conventional angiography,

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**Figure 4.** Three-dimensional virtual reality image showing anomalous origin of left circumflex artery (LCX) from right coronary artery (RCA; long white arrow), aorta (AO), and retroaortic course of LCX; short white arrow).

**Figure 5.** Multiplanar reconstruction image showing myocardial bridging of left anterior descending artery (arrow).

**Figure 6.** Three-dimensional virtual reality image showing myocardial bridging (arrow) of right coronary artery (RCA; white arrow).

**Figure 7.** Multiplanar reconstruction image showing hypoplastic (reduced calibre) left main coronary artery (white arrow) with normal ostium (black arrow).

Abbreviations: AO = aorta; LCS = left coronary sinus; LV = left ventricle.
nonselective injection into the left coronary sinus showed a hypoplastic LMCA and hypoplastic mid and distal LAD. The treatment of choice, coronary artery bypass grafting (CABG), was performed, with two grafts placed, one to the left internal mammary artery for the D1 branch and the other, a saphenous vein graft of the obtuse marginal artery.

Among other variants (Table 2), co-dominant circulation (n = 11; Figure 11), left dominant coronary circulation (n = 10; Figure 12), ramus intermedius (n = 15; Figures 9, 13 and 14), early origin of posterior descending artery (PDA, n = 5; Figure 15), and double PDA (n = 3) were noted.

DISCUSSION
Cardiac catheterisation has remained the gold standard imaging modality to visualise coronary anatomy. However, owing to the potentially complex three-dimensional nature of these anomalies, conventional coronary angiography incompletely delineates the anatomical course of the coronary arteries. The three-dimensional nature of multidetector CT coronary angiography datasets allows proper analysis of anomalous coronary arteries. We have presented a series of patients whose routine CTAs showed a wide range of anomalies in great detail, which could help the interventionalist and cardiac surgeon in the preoperative planning.
Figure 11. Three-dimensional virtual reality image showing co-dominant circulation. Posterior descending artery (PDA) arising from right coronary artery (RCA) and a posterolateral branch (PLB) arising from the left circumflex artery (LCX).
Abbreviations: AO = aorta; LAD = left anterior descending artery; LCA = left coronary artery; RCS = right coronary sinus.

Figure 12. Three-dimensional virtual reality image showing left-dominant circulation. Posterior descending artery (PDA) and posterolateral branches (PLB) arising from left circumflex artery (LCA; thick white arrow).
Abbreviations: AO = aorta; LAD = left anterior descending artery; LCS = left coronary sinus; OM = obtuse marginal artery.

Figure 13. Three-dimensional virtual reality image showing trifurcation of left coronary artery (LCA), ramus branch (thick white arrow), left anterior descending (LAD) artery, and left circumflex artery (LCX).
Abbreviations: AO = aorta; RCA = right coronary artery.

Figure 14. Three-dimensional virtual reality image showing trifurcation of left coronary artery (LCA), ramus branch (thick white arrow), left anterior descending (LAD) artery, left circumflex artery (LCX), and aorta (AO).
High Take-off of Right Coronary Artery

The positions of the coronary orifices are described in
terms of their relationship to the sinotubular junction.
We have reported a relatively less frequent variation
known as “high take-off” RCA.

High origin of coronary artery is defined as origin of a
coronary artery >1 cm above the sinotubular junction.4,5
According to studies based on autopsy examination,
high take-off anomaly with acute downward angulation
of the proximal RCA and acute downward angulation
of the LMCA led to sudden death.6 A few studies
identified infants with a high take-off of the RCA who
had coexisting ventricular septal defect and bicuspid
aortic valve. The importance of screening patients with
a bicuspid aortic valve for a high-origin RCA has also
been emphasised in the surgery literature.

The process of catheterisation of a coronary artery with
a high origin may be difficult. In patients with high
take-off of the RCA from anterior or especially left
anterior part of the ascending aorta, the right radial
approach is preferred over the femoral approach to
save time.7 Preoperative identification of a high-origin
coronary artery and its course are important in patients
undergoing aortotomy as part of aortic valve surgery or
ascending aortic replacement.

Cross-clamping of the aorta below a high-origin
coronary artery may result in unsuccessful induction of
cardioplegia. If the proximal part of the RCA is cut off,
it may lead to preoperative myocardial ischaemia. High
take-off RCA may also traverse the site of the planned
proximal saphenous vein graft anastomosis in CABG
surgery.

We documented two cases of high take-off RCA, of
which one was originating 11 mm above the sinotubular
junction, sharing its origin with the left coronary artery.
In one patient, the RCA also travelled an anomalous
interarterial course between the pulmonary artery and
aorta. Another high take-off RCA that has been identified
in our study originated from the ascending aorta above
the right coronary sinus showing normal course and
insertion.

Anomalous Right Coronary Ostium with
Interarterial Course

The anomalous origin of the RCA from the left coronary
sinus can be subdivided into two types based on the
location of the RCA ostium. In the high interarterial
course, the RCA ostium is located between the aorta
and the pulmonary artery. In this condition the proximal
segment of the anomalous RCA courses between the
aorta and pulmonary artery. This pattern of course is
called “malignant” as it is associated with sudden death
of the individual.8,9 In the low interarterial course, the
RCA ostium is located between the aorta and the right
ventricular outflow tract (RVOT). In this scenario, the
ostium is below the level of the pulmonary valve, with
no segment between the aorta and the RVOT.5

It is also known that a coronary artery arising from the
opposite sinus can take any of four common courses:
interarterial (between the aorta and the pulmonary artery),
retroaortic, prepulmonic, or subpulmonic (septal).

The possible mechanisms of restricted coronary blood
flow seen in interarterial courses are suggested to
be the acute take-off angle, a slit-like ostium, and
compression of the intramural segment by the aortic
valve commissure.5,10,11 The mechanism which may lead
to cardiac events in individuals with the high interarterial
subtype is that an ostium that is located above the
pulmonary valve would be compressed, due to the
blood that is forced into the aorta and pulmonary artery
during systole. This may also lead to compression of the
interarterial segment between them. Cases of sudden
death during exercise were reported. In cases with
coronary artery anomalies, the risk of death is higher in patients <30 years and the risk lower in aged people.3

However, in the low interarterial course variant, the RVOT contracts during systole, so an RCA ostium below the pulmonary valve would be less compressed between the aorta and RVOT. These coronary anomalies if detected in early stages of life and followed by proper treatment could prevent sudden death.

The choice of treatment according to most of the literature is surgical revascularisation in all cases. The available options are CABG and reimplantation of the coronary ostia and unroofing of the coronary artery. Unroofing of the coronary artery is being considered as a better treatment option if anatomically feasible.12,13 In Japan, the treatment for this condition is conservative with the patient being treated medically with beta blockers.12,14 In our study we report a case of anomalous origin of the RCA from the left coronary sinus showing an interarterial and malignant course between the pulmonary artery and aorta.

Left Circumflex Artery from Right Coronary Artery with Retroaortic Course

The ectopic origin of the LCX is considered the most common coronary anomaly and can be found in approximately 0.37% to 0.7% of all patients. The anomalous LCX most commonly arises from a separate ostium within the right coronary sinus, or as a proximal branch of the RCA.4 Although this anomaly was considered benign and asymptomatic, a few cases of sudden death, myocardial infarction, and angina pectoris in the absence of coronary artery disease have been reported.5

An anomalous origin of LCX from right coronary sinus is divided into three types: separate ostia for RCA and LCX (Type I), common ostia in the right sinus (Type II), or LCX arising as a branch of the proximal RCA (Type III).15

On selective conventional coronary angiography in the left anterior oblique and right anterior oblique projections the exact anatomical course is typically identified.16 Visualisation of the “dot” sign on the left ventricular angiogram in the right anterior oblique view (just posterior and to the left of the posterior aortic margin) is used as a clue by the coronary interventionalist for identification of an LCX arising from RCA with a retroaortic course.16 In our study we reported two cases of LCX originating from the right coronary sinus and showing a retroaortic course. Balloon angioplasty seems to be a favourable approach for revascularisation in these vessels.17-19 Appropriate selection of a guiding catheter is important.18

Coronary Hypoplasia

Hypoplastic coronary artery disease was first reported in 1970. It is underdevelopment of one or more major branches of the coronary arteries characterised by a narrowed lumen or shorter course.20-27 Its incidence is 0.02% of the general population and 2.2% of all the congenital coronary artery anomalies, however, its aetiology is still unknown. It was postulated to result from, for example, stenosis of the coronary artery orifice, an aberrant course between the pulmonary artery and aorta, a coronary artery ostium in ectopic position, or stenosis of the coronary ostium.9,23,24,27-31

The condition is mostly asymptomatic. However, some present with chest pain and palpitations. It bears a high risk of sudden cardiac death as a result of sudden and total occlusion of the artery. Mechanisms involved in cardiac events are coronary artery spasm reflecting abnormal vasodilator mechanisms and endothelial dysfunction leading to myocardial ischaemia. We reported a case of hypoplastic LMCA and LAD in a 33-year-old female as discussed in our results. This unusual clinical entity has rarely been diagnosed in living individuals. Most of them are asymptomatic and a high proportion experience sudden cardiac death. Diagnosis of this condition is often made at autopsy.

Myocardial Bridging of Coronary Arteries

Myocardial bridging of a coronary artery is defined as a band of myocardium overlying a segment of a coronary artery.32,33 It is most commonly localised in the middle segment of the LAD.

We documented myocardial bridging of LAD and RCA in our study; LAD was more commonly involved. Myocardial bridging in few cases might be responsible for angina pectoris, myocardial infarction, life-threatening arrhythmias, or even death. The typical “milking” effect and a “step down–step up” phenomenon induced by systolic compression of the tunnelled segment is considered as a standard for diagnosing myocardial bridging on conventional catheter angiography.4,33

Multidetector-row CTA clearly shows the intramyocardial location of the involved coronary
Coronary Artery Anomalies on CTA

artrial segment. The ECG-gated reconstruction window used in standard multidetector-row CTA of the coronary arteries is usually positioned within the diastolic phase for maximal vasodilatation and minimal motion artefacts. However, when there is suspicion for myocardial bridging, it is recommended that ECG-gated reconstruction be performed during the systolic phase as well as the diastolic phase. Comparison of the images obtained during the two phases will allow assessment of luminal narrowing during the systolic phase.

**Early Origin of Posterior Descending Artery**
According to previous studies in RCA-dominant individuals, 25% of them showed significant anatomical variations in the origin of the PDA.

The variations associated with PDA are: partial supply of the PDA territory by acute marginal branches, double PDA, and early origin of the PDA proximal to the crux. We reported five cases of early origin of PDA and three cases of double PDA in our study.

**Ramus Intermedius**
The most common variation in left coronary artery anatomy is the presence of a trifurcation of the LMCA. In this instance, the LMCA trifurcates into the LAD, LCX, and another artery called the ramus intermedius artery. The ramus intermedius artery has variable branching. It can be distributed as a diagonal branch if it supplies the anterior wall or as an obtuse marginal branch when it supplies the lateral wall.4,5

**Dominance**
The artery which supplies both the posterior descending branch and posterior left ventricular branch is considered dominant. Usually RCA gives both PDA and posterolateral branch.34 In few individuals both the branches arise from LCX, this is called left dominant coronary circulation.

There is another scenario in which RCA gives the PDA and LCX gives the posterolateral branch. This is stated to be co-dominant circulation. In approximately 70% of the population, the PDA originates from the RCA; it is co-dominant in 20% and 10% are left dominant.

According to a study by the National Cardiovascular Database Cath Percutaneous Coronary Intervention Registry, left coronary dominance was associated with 1.19-fold increased odds of in-hospital mortality, and co-dominance was associated with 1.16-fold increased odds of death, after percutaneous coronary intervention for acute coronary syndrome after accounting for 23 demographic, clinical, and angiographic characteristics.35

The detailed evaluation and reporting of the anomalies and variants in the coronary tree is essential, primarily as it might be the cause of the patient’s symptoms and it guides the surgeon and/or interventionalist during cardiac procedures, thereby reducing the intra- and post-operative events.

**CONCLUSION**
Coronary artery anomalies occur in 1% to 5% of patients undergoing coronary arteriography. Despite the large number of reported anomalies, congenital anomalies of coronary arteries are present in <3% of all congenital heart diseases, and <1% among the general population.4

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