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Anomalies of coronary artery origin: Evaluation on multidetector CT angiography

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ABSTRACT

Anomalies of origin of coronary arteries are an uncommon occurrence and found in approximately 1–2% of the general population. While a large proportion of these anomalies are clinically silent, a few might be hemodynamically significant and may even result in sudden cardiac death. Comprehensive knowledge of the normal as well as variant anatomies of the coronary artery origin and familiarity with imaging appearances and clinical significance of these anomalies is imperative for precise diagnosis and subsequent planning of treatment, whenever required. Multidetector computed tomography angiography, on account of its non-invasiveness, faster scan times and multiplanar reconstruction capabilities, is increasingly being utilized for characterization of coronary artery origin anomalies and their three-dimensional spatial relations. It shows a superior rate of detection of these anomalies compared to conventional angiography, providing more accurate delineation of the ostium as well as course. With the advent of newer generation CT scanners and use of advanced dose reduction techniques, images can be obtained rapidly having excellent spatial resolution and with minimal radiation dose. In this review article, we present the multidetector CT angiography imaging findings of the spectrum of anomalous coronary artery origin, using a third-generation dual-source CT scanner.

1. Introduction

Anomalies of origin of coronary arteries are rare and seen in approximately 1–2% of the general population [1]. Although uncommon, they can be hemodynamically significant and can even result in sudden cardiac death (SCD). Comprehensive knowledge of the normal as well as variant anatomies of the coronary artery origin is imperative for the diagnosis and planning possible treatment, if required. Multidetector computed tomography (MDCT) angiography is increasingly being utilized for characterization of coronary artery origin anomalies and their three-dimensional spatial relations, due to its non-invasiveness, faster scan times and multiplanar reconstruction capabilities. With the advent of newer generation CT scanners and use of advanced dose reduction techniques, images can be obtained rapidly having excellent spatial resolution and with reduced radiation dose.

2. MDCT angiography technique

All examinations were performed on a performed on a SOMATOM FORCE (Siemens Healthcare, Forchheim, Germany) CT scanner. The scanner is a third generation dual source CT scanner with a temporal resolution of as low as 66 milliseconds. Along with increasing the speed of acquisition, the use of advanced dose reduction techniques also help reduce the radiation dose substantially.

Retrospective ECG-gated CT angiography examination was performed after injection of non-ionic iodinated contrast (Omnipaque 350, GE Healthcare, Princeton, NJ, USA; 1.0 mL/kg body weight) via a peripheral intravenous line using a dual head power injector at a flow rate of 4.5 mL/s followed by a 50-mL saline chaser injected at the same flow rate. A 'bolus tracking' method was used whereby CT acquisition was automatically triggered when contrast opacification threshold of 100 Hounsfield units (HU) was achieved in the ascending aorta on the monitoring sequence. Automated tube voltage selection and automated tube current modulation based on body habitus (CARE kV and CARE Dose4D, Siemens Healthcare) were enabled, with available tube voltage settings ranging from 70 to 120 kV in 10 kV increments.

Slices were reconstructed of 0.6-mm section thickness and increment of 0.4 mm, using a medium sharp kernel (Bv40), with a model-

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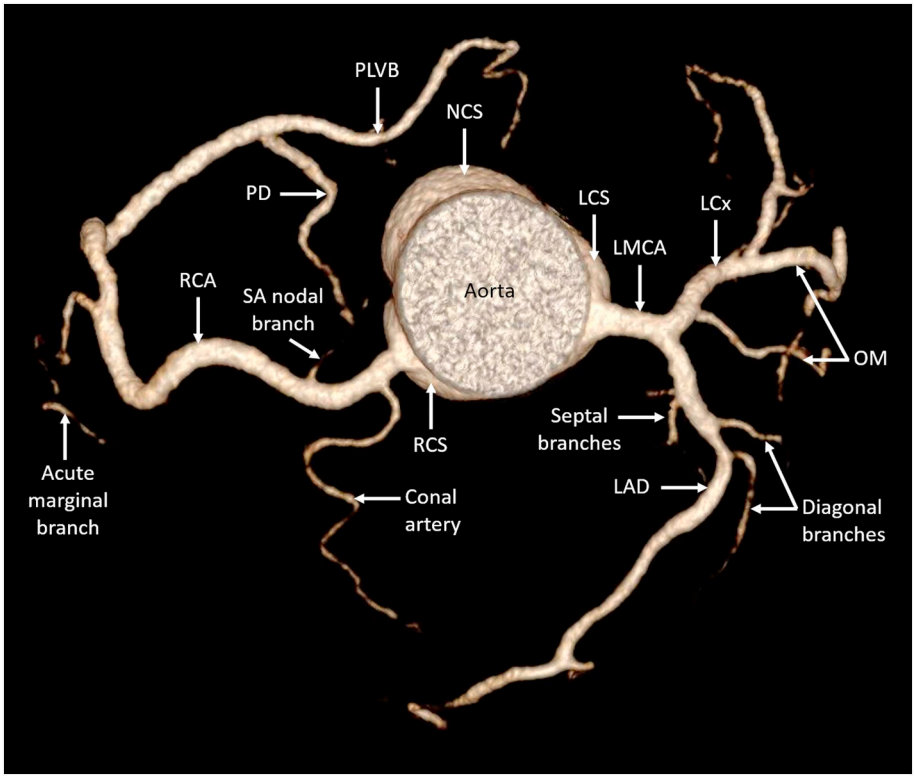

Fig. 1. Normal origin of coronary arteries.
Volume rendered image shows right coronary artery (RCA) arising from the right coronary sinus (RCS) of Valsalva while the left main coronary artery (LMCA) arising from the left coronary sinus (LCS) of Valsalva. The non-coronary sinus (NCS) gives off no coronary artery. (PD: posterior descending artery; PLVB: posterior left ventricular branch; LAD: left anterior descending artery; LCx: left circumflex artery; OM: obtuse marginal branches; SA: sino-atrial.)

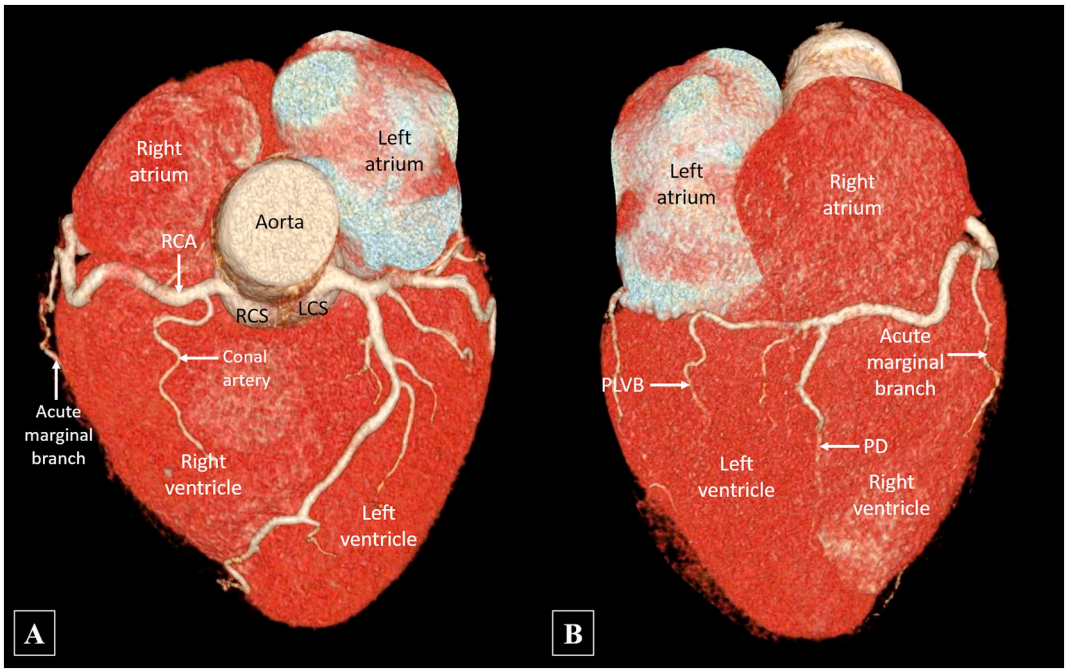

Fig. 2. Normal course and branching of the right coronary artery.
Volume rendered images (A and B) depict the right coronary artery (RCA) coursing in the right atrioventricular groove and giving off the conal artery and the acute marginal branch and bifurcating into the posterior descending (PD) artery and posterior left ventricular branch (PLVB). (RCS: right coronary sinus; LCS: left coronary sinus.)

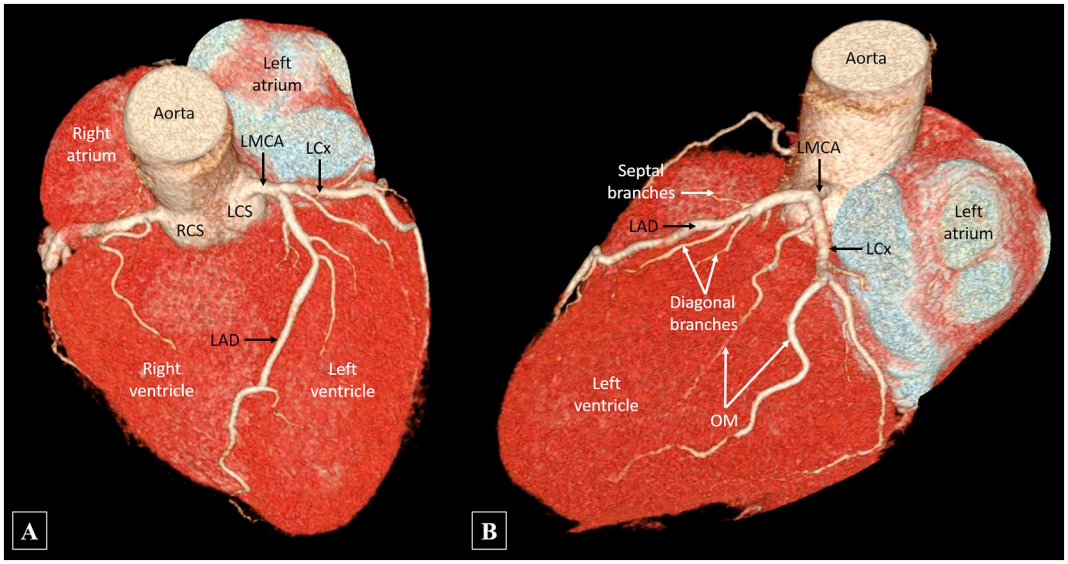

Fig. 3. Normal course and branching of the left coronary artery.
Volume rendered images (A and B) depict the left main coronary artery (LMCA) bifurcating into the left anterior descending (LAD) artery and the left circumflex (LCx) artery. The LAD artery is seen coursing in the anterior interventricular groove giving off septal and diagonal branches while the LCx artery enters the left atrioventricular groove and gives off obtuse marginal (OM) branches. (RCS: right coronary sinus; LCS: left coronary sinus.)

Table 1
Classification of anomalies of coronary artery origins.

Type 1	Coronary stenosis or atresia
Type 2	Anomalous origin of coronary artery from aorta with/without anomalous course
A	High take off
B	Multiple ostia
C	Single coronary artery
D	Origin of a coronary artery from the opposite or non-coronary sinus of Valsalva
	i. Right coronary artery from left coronary sinus (or left main coronary artery/left anterior descending artery)
	ii. Left main coronary artery from right coronary sinus (or right coronary artery)
	iii. Left circumflex artery or left anterior descending artery arising from the right coronary sinus (or right coronary artery)
	iv. Right or left main coronary artery (or a branch of either artery) arising from the non-coronary sinus
Type 3	Anomalous origin of coronary artery from pulmonary artery
	i. Left main coronary artery from pulmonary artery (ALCAPA)
	ii. Right coronary artery from pulmonary artery (ARCAPA)
	iii. Accessory coronary artery from pulmonary artery
	iv. Entire coronary circulation from pulmonary artery
Type 4	Extra-aortic and extra-pulmonic origin of coronary artery

based iterative reconstruction strength level 3 (ADMIRE; Siemens Healthcare). Axial sections were analyzed along with coronal, sagittal and curved multiplanar reformats (MPR), followed by volume rendered (VR) and maximum intensity projection (MIP) images on an external workstation (Multi-Modality Workplace; Siemens Healthcare).

3. Normal origin of coronary arteries

The right coronary artery (RCA) arises from the anterior right coronary sinus (RCS) of Valsalva at a slightly inferior level compared to origin of the left main coronary artery (LMCA). The RCA courses in the right atrioventricular groove and gives off the conal artery as its first branch, unless it has a separate origin from the RCS. The sino-nodal branch is usually the second branch of the RCA, although in a small percentage of patients it may arise off the left circumflex artery. The other branches include the acute marginal branch, two or three right ventricular branches and in a right dominant circulation, the posterior descending (PD) artery and posterior left ventricular (PLV) branch (Figs. 1 and 2) [2].

The LMCA arises from the left coronary sinus (LCS) of Valsalva and usually bifurcates into the left anterior descending (LAD) artery and the left circumflex (LCx) artery or can trifurcate giving off an additional ramus intermedius (RI) branch. The LAD artery courses in the anterior interventricular groove and gives off septal and diagonal branches. The LCx artery enters the left atrioventricular groove and gives off obtuse marginal branches. In left dominant circulation, it supplies the PD artery and PLV branch as well (Figs. 1 and 3) [2].

4. Anomalies of the origin of coronary arteries

Varying classification systems have been proposed by different authors in an attempt to encompass the multitude of coronary arterial origin anomalies [3]. However, a simplistic yet exhaustive classification of anomalies of coronary artery origin remains elusive. In this article, these anomalies have been classified into four major categories including coronary stenosis or atresia, anomalous origin of coronary artery from aorta with/without anomalous course, anomalous origin of coronary artery from pulmonary artery and extra-aortic/extra-pulmonic origin of coronary artery (Table 1).

4.1. Type 1: coronary stenosis or atresia (COSA)

COSA is a spectrum of coronary anomalies which share two common features; the anomaly being congenital in origin and the defect resulting in ostial or proximal coronary stenosis or atresia. Though the

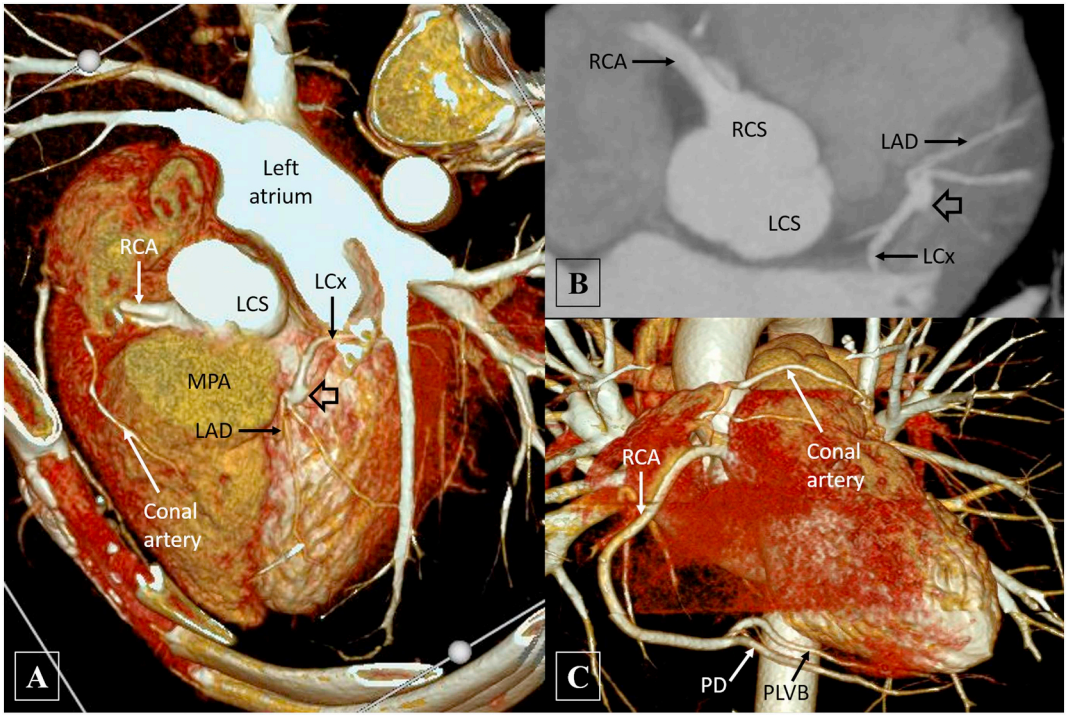

Fig. 4. Left main coronary artery atresia.
Volume rendered images (A and C) and maximum intensity projection oblique axial image (B), shows absence of the left main coronary artery with reformation (block arrow) of the left anterior descending (LAD) artery and the left circumflex (LCx) artery. No coronary artery is seen originating from the left coronary sinus (LCS). It also shows normal origin of right coronary artery (RCA) from the right coronary sinus (RCS) with hypertrophied conal artery along with hypertrophied posterior descending (PD) artery and posterior left ventricular branch (PLVB). (MPA: main pulmonary artery.)

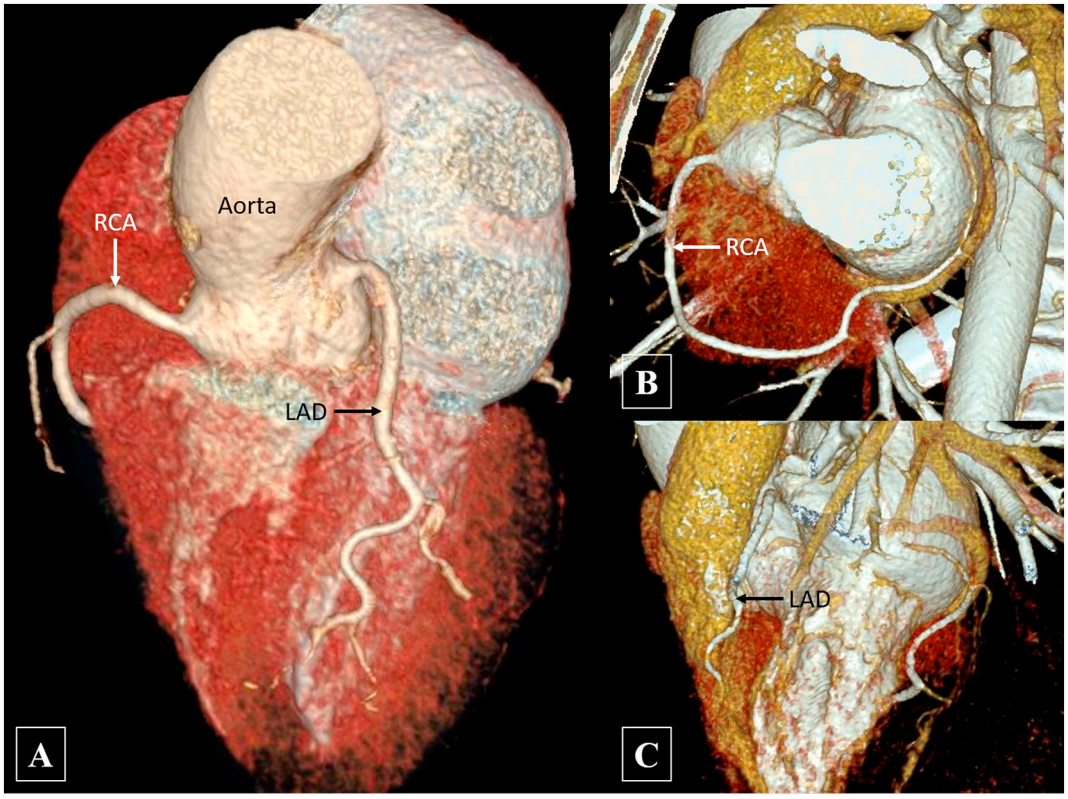

Fig. 5. Left circumflex artery atresia.
Volume rendered images (A, B and C) shows continuation of the left main coronary artery as the left anterior descending (LAD) artery with absence of left circumflex (LCx) artery in the left atrioventricular groove. A super-dominant right coronary artery (RCA) is seen reaching the left atrioventricular groove.

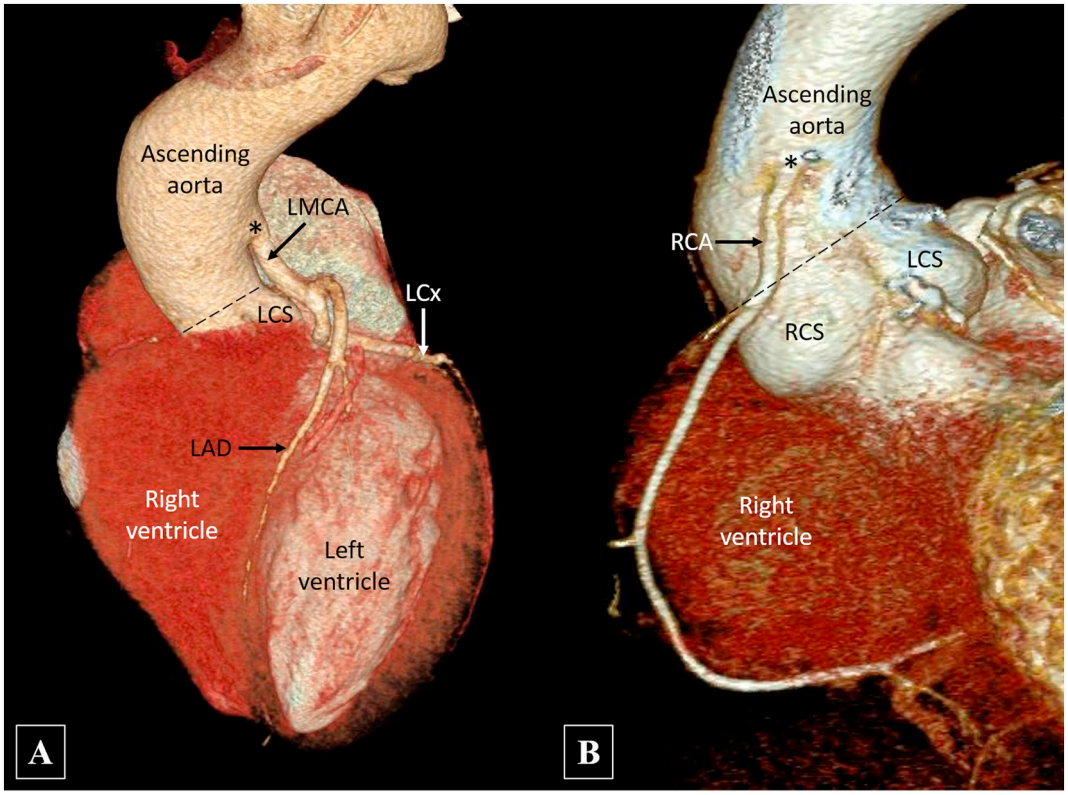

Fig. 6. High take-off of coronary arteries.
Volume rendered images (A and B) reveal high take-off (*) of the left main coronary artery (LMCA) and right coronary artery (RCA) respectively, from the ascending aorta, well above the sino-tubular junction (dotted line). (RCS: right coronary sinus; LCS: left coronary sinus; LAD: left anterior descending artery; LCx: left circumflex artery.)

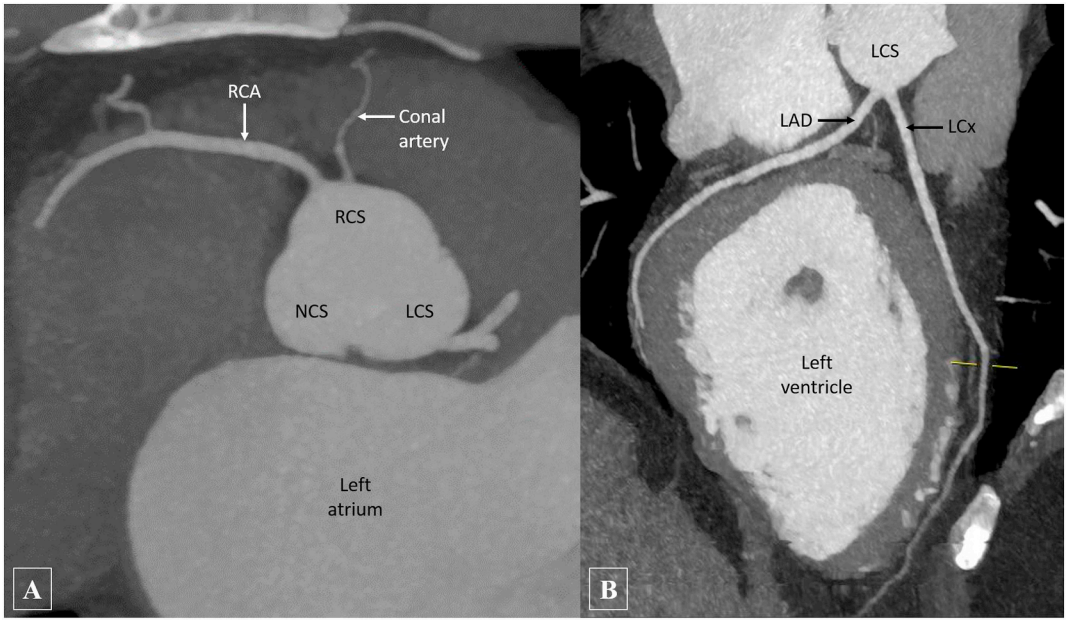

Fig. 7. Multiple ostia.
Axial maximum intensity projection image (A) shows direct origin of the conal artery from the right coronary sinus (RCS), separate from the origin of the right coronary artery (RCA). Curved multiplanar image (B) of a different patient, shows absence of the left main coronary artery with direct separate origins of the left anterior descending (LAD) artery and the left circumflex (LCx) artery from the left coronary sinus (LCS); however, the ostia are very close to each other. (NCS: non-coronary sinus.)

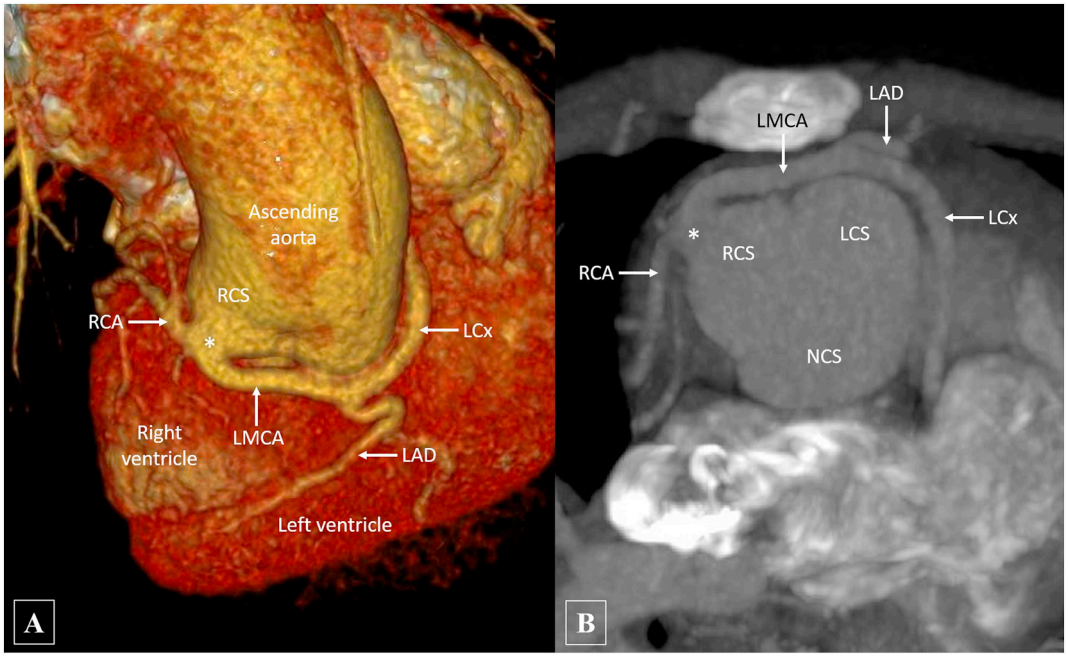

Fig. 8. Single coronary artery.
Volume rendered image (A) and maximum intensity projection oblique axial image (B), show a single coronary artery arising (*) from the right coronary sinus (RCS) and immediately bifurcating into the right coronary artery (RCA) and the left main coronary artery (LMCA). The LMCA further branches into the left anterior descending (LAD) artery and the left circumflex (LCx) artery. All the three coronary arteries (RCA, LAD and LCx) then course in their normal expected locations. (NCS: non-coronary sinus; LCS: left coronary sinus.)

defect is always congenital, it might be asymptomatic at birth owing to mild non-significant stenosis [4]. With time, this stenosis may progress in severity or proceed to a total occlusion, thus becoming symptomatic. In adult patients with suspicion of COSA, it is imperative to rule out acquired causes such as atherosclerosis and arteritis first.

COSA involves the LMCA more commonly than the RCA; the coronary ostium of which can be normally located or be ectopic (Figs. 4 and 5). Some cases of COSA can be recognized by the presence of an ostial dimple and a blind distal left main trunk. Also, the presence of only one or two full-diameter non-stenotic connecting collateral vessels is indicative of the congenital origin of this defect. Myocardial scarring is rare in COSA and only rarely is a surgical intervention required, to bypass the stenosis or atresia, owing to the presence of a good sized collateral network.

4.2. Type 2: anomalous origin of coronary artery from aorta with/without anomalous course

4.2.1. High take off

It indicates to the origin of either the RCA or LMCA at a point more than 5 mm above the sino-tubular junction, from the ascending aorta (Fig. 6). It is more commonly seen with the RCA with the prevalence of bilateral high take off reported to be up to 6% [5]. The artery may also have an intramural course. This variant usually has no inherent hemodynamic significance, but may pose difficulty in cannulating the vessels during coronary arteriography [6]. Also, cross-clamping the aorta below the high origin coronary artery can lead to unsuccessful cardioplegia during surgeries such as aortic valve replacement. In root sparing aortic graft repair, this may pose an added difficulty.

4.2.2. Multiple ostia

Typically, there is separate origin of the RCA and its conal branch from the RCS or the LAD and LCx arteries arise separately from the LCS with no LMCA (Fig. 7). Again, these may result in difficulties in cannulation at arteriography. A separate conal artery may also be at risk for injury during procedures like ventriculostomy performed during congenital heart surgery [7].

4.2.3. Single coronary artery

This is an extremely rare congenital anomaly with a single coronary ostium from the aortic trunk which either follows the pattern of a normal RCA/LMCA or divides into two branches with distributions of the RCA and LMCA respectively (Fig. 8), or has a distribution entirely different from that of the normal coronary arterial tree. The two arteries can either share a proximal common trunk with an ectopic origin of one of the arteries from the opposite sinus or one of the arteries can originate from a distal segment of the contralateral artery. These patients are at risk for SCD if a major coronary branch has an interarterial course. Also, a single coronary artery developing a proximal stenosis in the absence of collateralization can be fatal [8].

4.2.4. Origin of a coronary artery from the opposite or non-coronary sinus of Valsalva

This may relate to arising from the contralateral or non-coronary cusp, sharing a common ostium with the contralateral coronary artery, or arising directly from the contralateral artery. There are four major patterns which include RCA arising from LCS, LMCA arising from RCS, LCx or LAD artery arising from RCS, and the LMCA or RCA arising from the non-coronary sinus (Figs. 9, 10 and 11). The anomalous artery can traverse one of the following four common courses, for example, interarterial (between the aorta and pulmonary artery; carries a high risk

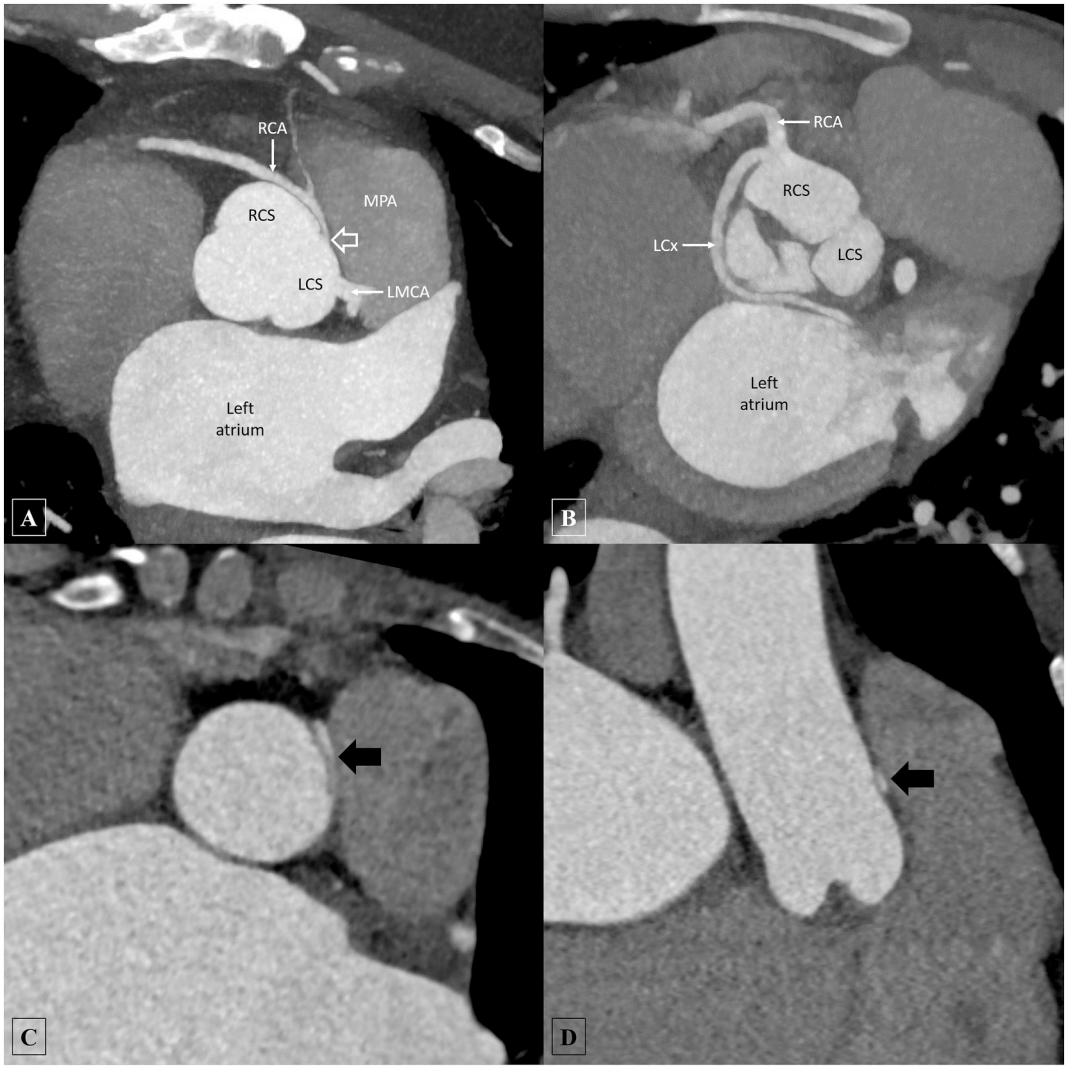

Fig. 9. Origin of a coronary artery from the opposite sinus of Valsalva.
Axial maximum intensity projection image (A) shows origin of right coronary artery (RCA) from the left coronary sinus (LCS) with a narrowed ostium (block arrow) and coursing between the aorta and main pulmonary artery (MPA). Axial maximum intensity projection image (B) of a different patient shows origin of the left circumflex (LCx) artery from the right coronary sinus (RCS), which then courses posterior to the aorta to enter the left atrioventricular groove. Axial (C) and oblique coronal (D) maximum intensity projection images of another patient reveal the elongated intramural course (resulting in a slit-like appearance) of the right coronary artery (block arrow), having an anomalous origin from the left coronary sinus. (LMCA: left main coronary artery.)

of SCD), retroaortic, prepulmonic, or trans-septal (subpulmonic). Table 2 summarizes the incidence of the various patterns of coronary origin from the opposite or non-coronary sinus with their common courses, risk of SCD and major associations.

The interarterial course can be either high or low depending on its relative position with respect to the pulmonary valve. The high interarterial course is termed as malignant course on account of compression of the stenotic/slit-like ostium that occurs during simultaneous distension of the aorta and pulmonary artery at systole, which predisposes the individual to SCD. However, without the presence of enlarged pulmonary arteries or pulmonary arterial hypertension, it is unlikely that the pulmonary artery would exert enough pressure to occlude the interarterial segment of the anomalous coronary artery [9]. Other additional morphological features which contribute to the risk of SCD include an acute angle take-off from the aorta and a course within the wall of the aorta (termed as intramural interarterial) which can result in a slit-like coronary artery ostium (Fig. 9C and D). With expansion of the aorta during exercise, further stretching of the intramural segment of the coronary artery may occur with consequent flap-like occlusion of the segment and myocardial ischemia [9–12].

A retroaortic course is usually seen when the LCx artery arises from the RCS and courses posterior to the aorta. This course usually does not have hemodynamic consequence, but may complicate aortic valve surgery. A prepulmonic course (most commonly LMCA arising from the RCS) indicates a course anterior to the right ventricular outflow tract (RVOT) and usually does not have any hemodynamic consequence; although can complicate corrective surgery for repair of RVOT stenosis in tetralogy of Fallot. A trans-septal (subpulmonic) course is differentiated from interarterial course as the artery does not show a slit like ostium and is surrounded by septal myocardium at some point. In the

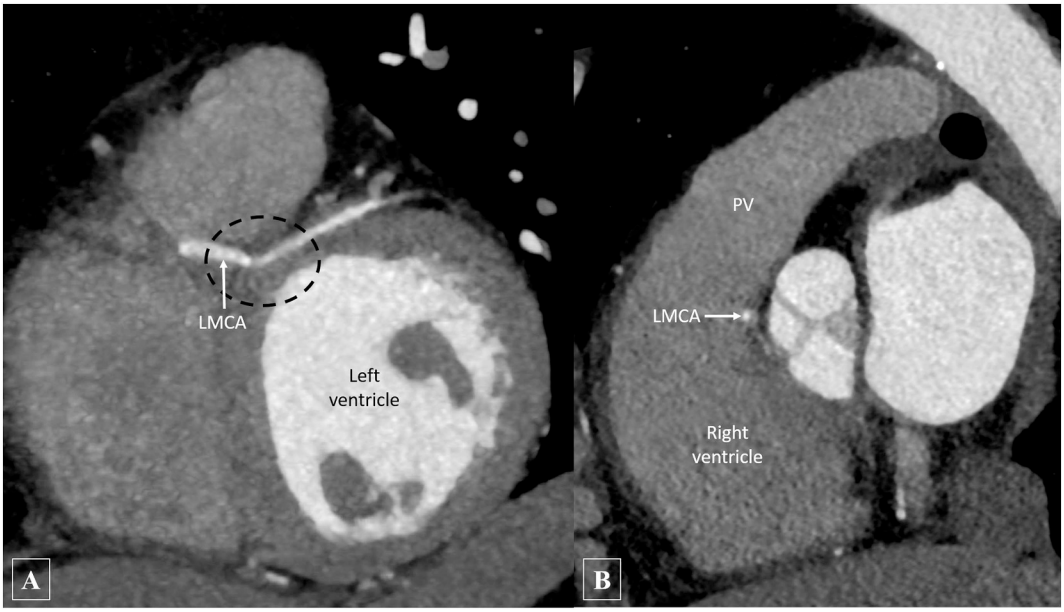

Fig. 10. Origin of a coronary artery from the opposite sinus of Valsalva with transseptal course.
Oblique coronal (A) and oblique sagittal (B) maximum intensity projection images demonstrate the hammock-like descending curvature (dotted circle) of the elongated left main coronary artery (LMCA), having an anomalous origin from the right coronary sinus. This is consistent with a transseptal course, which is located lower than expected in interarterial course in the sagittal plane. (PV: pulmonary valve.)

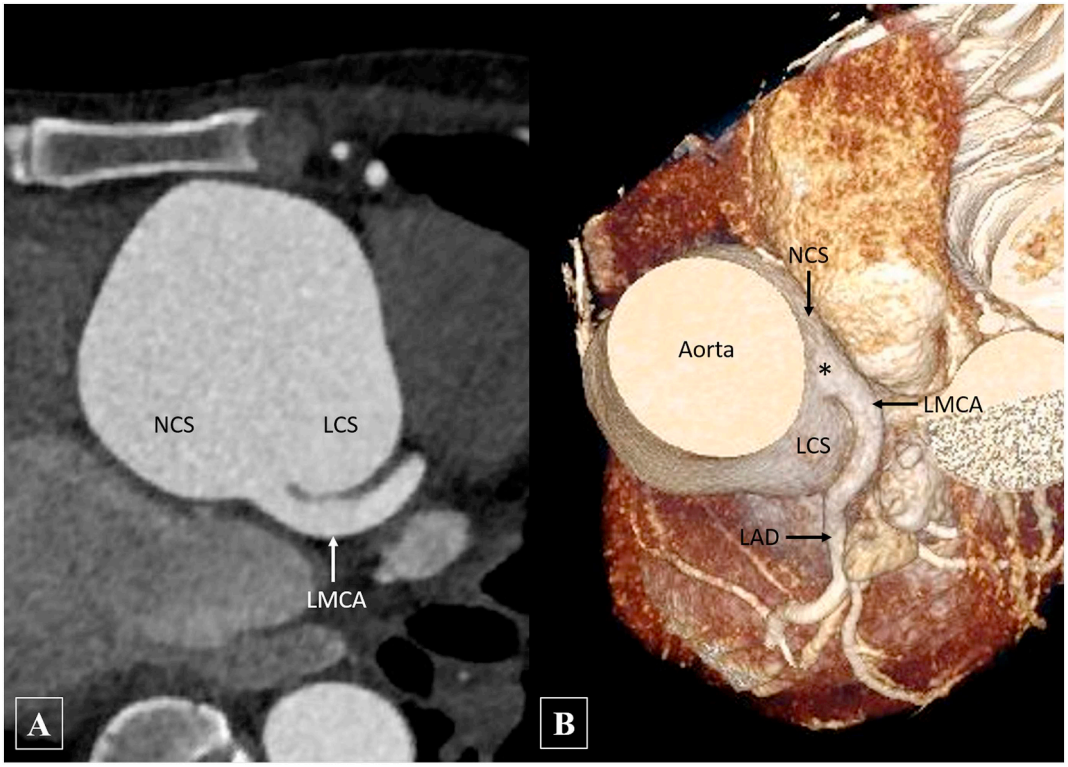

Fig. 11. Origin of a coronary artery from the non-coronary sinus of Valsalva.
Axial maximum intensity projection image (A) and volume rendered image (B) reveals origin (*) of the left main coronary artery (LMCA) from the non-coronary sinus (NCS) close to the commissure. No coronary artery is seen originating from the left coronary sinus (LCS). (LAD: left anterior descending artery.)

Table 2
Incidence of various patterns of coronary origin from the opposite sinus with their common courses, risk of sudden cardiac death and important associations [8,15].

Anomaly	Incidence	Course	Risk of sudden cardiac death	Clinical considerations	Important associations
RCA from LCS (or from LMCA/LAD artery)	Up to 0.9% in patients undergoing angiography	**Most common - interarterial** "High" interarterial: anomalous ostium located between Ao and MPA - simultaneous distension of Ao and MPA during systole compresses the ostium. "Low" interarterial: anomalous ostium located between Ao and RVOT - Less compression of the ostium as distension of aorta is accompanied by simultaneous contraction of RVOT during systole. **May have intramural course:** Shows a slit like course with lack of fat plane between vessel and Ao. Increase in caliber is noted most distally (usually about 0.5–1.0 cm) where it emerges from the wall.	SCD with interarterial and intramural course	The importance of identifying intramural course lies in the different surgical approach for an intramural RCA (i.e. unroofing) vs re-implantation	
LMCA from RCS (or RCA)	6–10 times less common than RCA from LCS	An interarterial course may be seen in up to 75% of patients. Retroaortic, prepulmonic and subpulmonic course also seen.	SCD with interarterial course: more common in anomalous LMCA than anomalous RCA with interarterial course Prepulmonic course: no hemodynamic consequences	Prepulmonic course seen in conotruncal anomalies may complicate surgical approach	Seen in conotruncal anomalies
LCx from RCS (or RCA)	Most common anomaly in this group: arises from a separate ostium within the RCS or as proximal branch of the RCA	Invariably takes a posterior course behind the aorta	Not associated with SCD	Risk of injury during aortic valve surgeries	
LAD artery from RCS (or RCA)	Rare in patients without CHD	Interarterial or prepulmonic course	SCD with interarterial course		Seen in TOF, DORV and TGA
RCA or LMCA from non-coronary sinus	Rare in patients without CHD	Posterior course	No hemodynamic consequences		May be seen in TGA

Ao: aorta; MPA: main pulmonary artery; RCA: right coronary artery; LMCA: left main coronary artery; LCx: left circumflex; LAD: left anterior descending; RCS: right coronary sinus; LCS: left coronary sinus; SCD: sudden cardiac death; TOF: tetralogy of Fallot; DORV: double outlet right ventricle; CHD: congenital heart disease; TGA: transposition of great arteries.

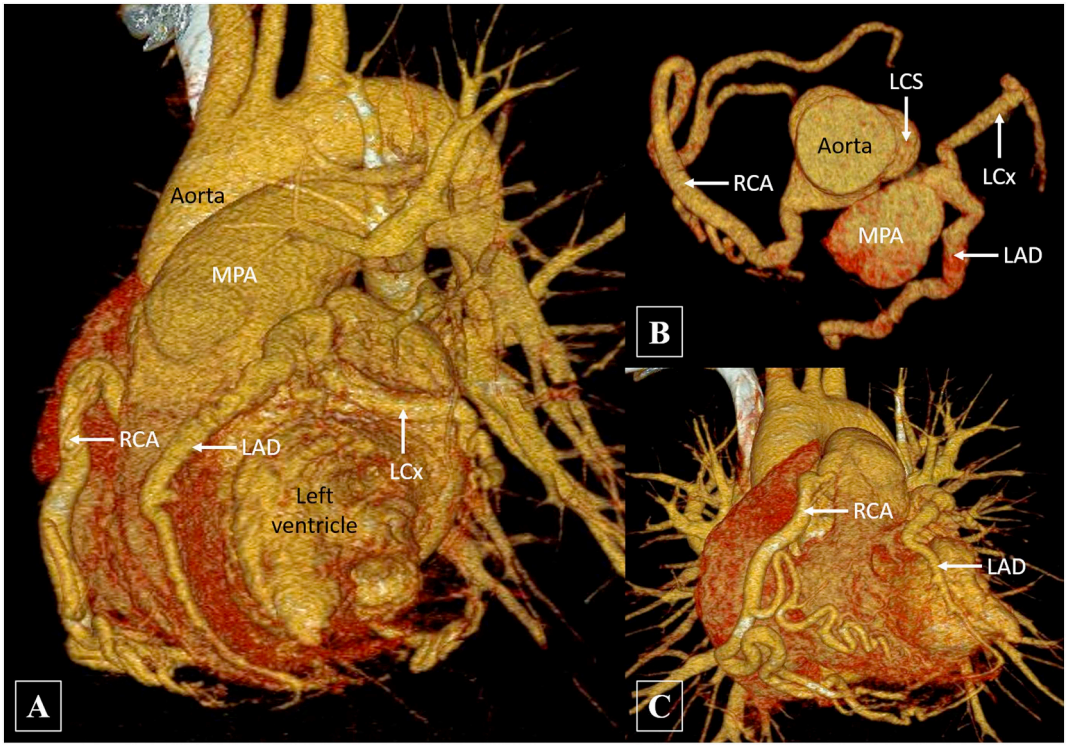

Fig. 12. Anomalous origin of left main coronary artery from pulmonary artery.
Volume rendered images (A, B and C) show anomalous origin of the left main coronary artery (LMCA) from the main pulmonary artery (MPA) with no coronary artery originating from the left coronary sinus (LCS). The right coronary artery (RCA) and its branches are hypertrophied and seen anastomosing with the branches of left anterior descending artery (LAD). (LCx: left circumflex artery.)

sagittal plane, it is located lower than expected in interarterial course while in the short axis plane, it is seen caudal to the crista supraventricularis.

4.3. Type 3: anomalous origin of coronary artery from pulmonary artery (ACAPA)

There are four patterns of ACAPA including, origin of LMCA from the pulmonary artery (ALCAPA), origin of the RCA from pulmonary artery (ARCAPA), origin of an accessory coronary artery from the pulmonary artery and origin of the entire coronary circulation from the pulmonary artery. ACAPA is not associated with any syndromes or non-cardiac conditions and is not considered an inheritable congenital cardiac defect.

ALCAPA with normal origin of RCA is known as Bland-White-Garland syndrome and is confirmed on coronary angiography which also demonstrates collateral circulation between the RCA and left coronary artery and a coronary “steal” phenomenon (Fig. 12) [13]. ALCAPA is more common than ARCAPA and is usually symptomatic in the first year of life, with 90% of the affected babies dying in the first year of their life due to coronary steal phenomenon [14]. In adults, it can be silent or can present with myocardial infarction, ischemic ventricular dysfunction, arrhythmias and SCD. ARCAPA is relatively uncommon and in a review by Radke et al., only 41% of ARCAPA were symptomatic and 40% had documented ischemia [15]. Other extremely rare variations include anomalous origin of LAD or LCx arteries from the pulmonary artery. Anomalous origin of the LCx from the pulmonary artery is usually discovered in childhood (Fig. 13). The presence of this defect in adults without the presence of congenital cardiac defects is extremely rare. Table 3 summarizes the associations of various patterns of ACAPA.

4.4. Type 4: extra-aortic and extra-pulmonic origin of coronary artery

Unusual anomalous origins of coronary arteries from the brachiocephalic trunk, left internal mammary artery, left subclavian artery, carotid artery and the bronchial artery have also been reported [3].

5. Conclusion

A thorough knowledge of the imaging manifestation of coronary origin anomalies is imperative for precise diagnosis and subsequent planning of treatment, whenever required. While a large proportion of these anomalies are clinically silent, a few might be hemodynamically significant and may even result in sudden cardiac death. Multidetector CT angiography with its multiplanar reconstruction capabilities and volume rendering techniques, has a superior rate of detection of these anomalies compared to conventional angiography providing more accurate delineation of the ostium as well as the course. Thus, having an awareness regarding the appearance of these anomalies on CT angiography and an insight into their clinical significance must be stressed upon.

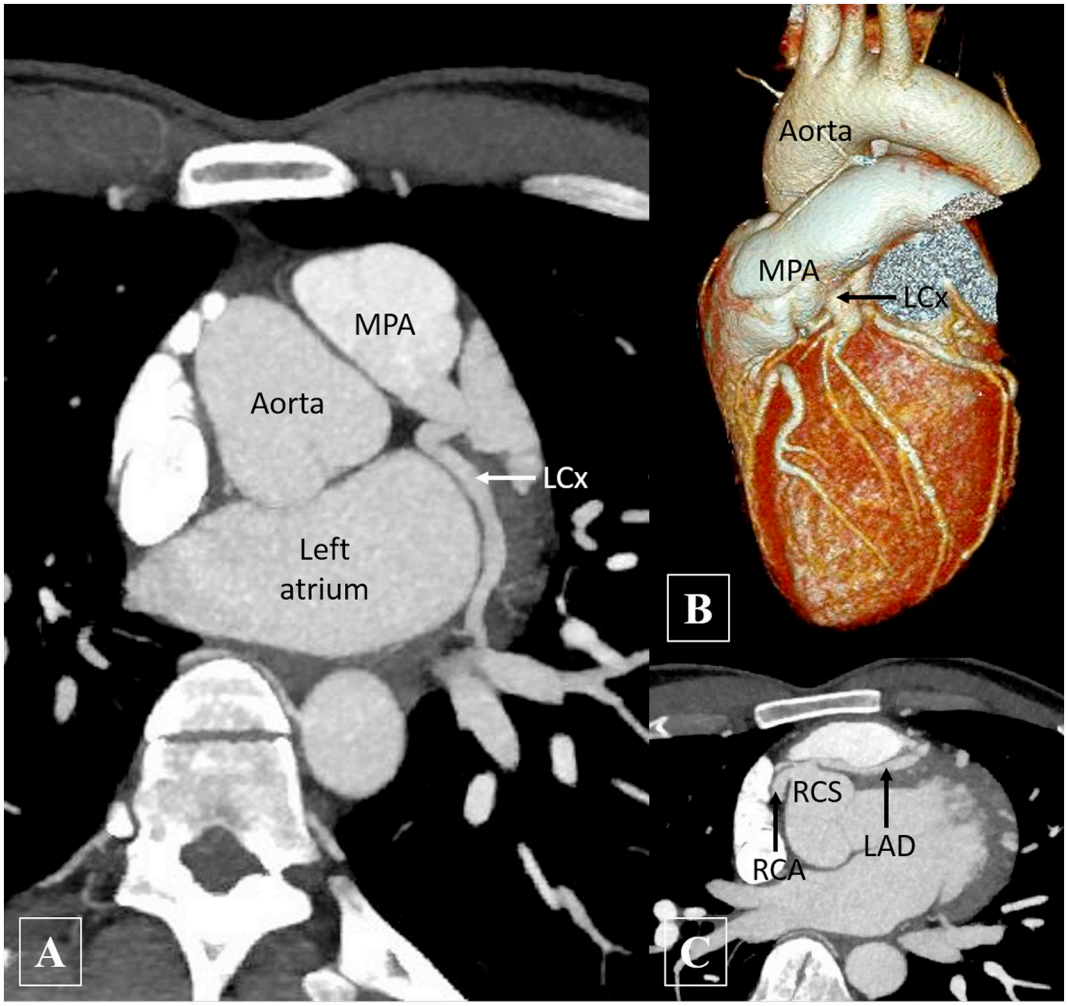

Fig. 13. Anomalous origin of left circumflex artery from pulmonary artery.
Axial maximum intensity projection images (A and C) and volume rendered image (B) reveal anomalous origin of the left circumflex artery (LCx) from the main pulmonary artery (MPA) with no coronary artery originating from the left coronary sinus. The right coronary artery (RCA) and left anterior descending artery (LAD) are seen to arise from the right coronary sinus (RCS).

Table 3
Incidence and associations of various patterns of anomalous origin of coronary artery from pulmonary artery.

Anomaly	Associations
ALCAPA	Patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot, or coarctation of aorta, rarely may be associated
ARCAPA	Aorto-pulmonary window, tetralogy of Fallot and septal defects are seen in 1/3rd of the patients with ARCAPA
Anomalous LCx from RPA	Most of the cases reported in the literature are associated with aortic arch anomalies and aortic coarctation and seen in conjunction with other congenital malformations such as tetralogy of Fallot, aorto-pulmonary window, patent ductus arteriosus, pulmonary valve stenosis, sub-aortic fibrous membrane stenosis

ALCAPA: anomalous origin of left coronary artery from the pulmonary artery; ARCAPA: anomalous origin of right coronary artery from the pulmonary artery; LCx: left circumflex; RPA: right pulmonary artery.

Declaration of Competing Interest

The authors declare no conflicts of interest.

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