

Normal Anatomy and Congenital Abnormalities of the Coronary Arteries

2

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2.1 Introduction

An adequate knowledge of the anatomy of coronary arteries and its normal variants is an important point for the analysis of MDCT images. Nomenclature of coronary anatomy is frequently confusing, as a number of anatomical, clinical and radiological terms are used in combination. This conventional terminology is useful, however, as it has been applied in conventional coronary angiography¹, and it will be maintained in the present chapter.

The heart is a highly differentiate blood vessel, with developed muscular walls. The vascular nutrition of myocardium is complex, with a number of anatomical normal variants that can involve even extracardiac vessels, such as bronchial, mammary and mediastinal arteries^{2,3}.

The coronary arteries are conductive vessels running through the epicardial surface of the heart, embedded in adipose tissue, and showing short segments of mild penetration into the myocardial tissue³.

As indicated by its name (from the latin *corona*: wreath), coronary arteries are distributed over the heart as a crown-shape network, showing anastomotic communications between its different branches, particularly at the level of the base and the apex of the left ventricle. The connection between divisions of the same artery is known as homocollateral circulation, and the connection between different arteries is named heterocollateral circulation. Physiological collateral circulation acquires a relevant role in pathological circumstances².

Macroscopical appearance of coronary arteries is variable in terms of diameter, which is larger in the left artery than in the right one in more than half of individuals, while the opposite occurs in nearly 20%². Also, the number of ramifications, its course—linear or sinusoid—(Figure 2.1), and the distance from the epicardial surface (Figure 2.2) is variable between individuals³.

Coronary arteries emerge from the aorta through the coronary *ostia*, located at the right (or anterior) and the left (or left posterior) sinuses of Valsalva². The coronary *ostia* (Figure 2.3) are situated at the level of the sinotubular junction or slightly below it (56% of cases), followed by a high left orifice and a low right orifice or at the level of the junction (30% of individuals)⁴.



FIGURE 2.1

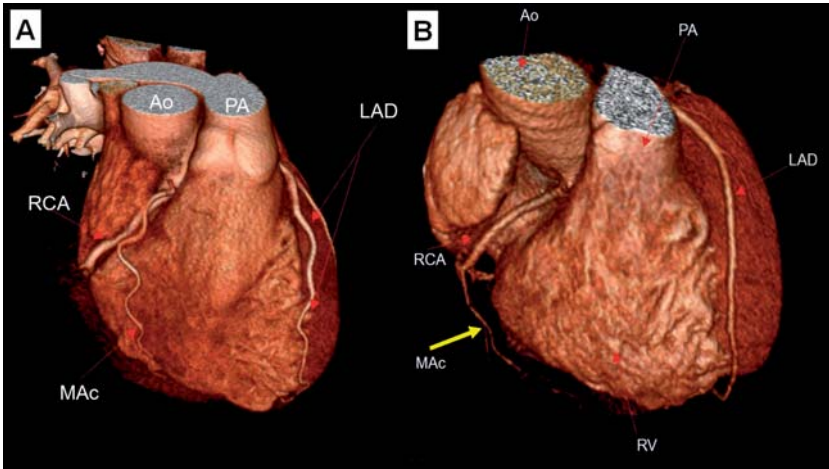


FIGURE 2.2

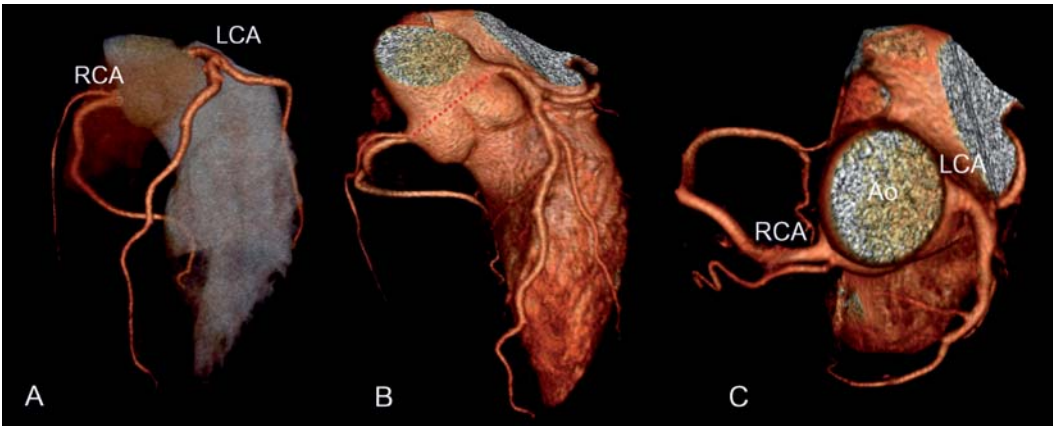


FIGURE 2.3

- F. 2.1. Coronary arteries of tortuous course (arrows): this normal variant is frequently found in hypertensive patients.
- F. 2.2. Differing distance of coronary arteries from epicardium: in patient A, the marginal acute (MAc) branch of the right coronary artery (RCA) courses close to the epicardium, while in patient B it runs at a distance from the external surface of the heart (arrow), this space being occupied by fat (deleted by filtering of the image). Ao: aorta; LAD: left anterior descending; PA: pulmonary artery; RV: right ventricle.
- F. 2.3. Origin of the coronary arteries as seen from frontal (A and B) and cranial (C) views: observe that the right coronary *ostium* is anterior and caudal with respect to the left one, while both are equidistant from the coaptation point of the aortic valve (B panel). Ao: aorta; LCA: left coronary artery; RCA: right coronary artery.

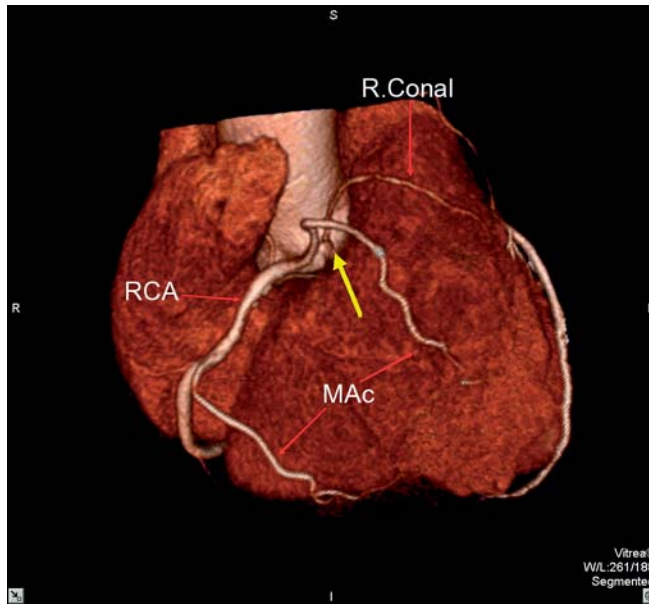


FIGURE 2.4 Independent origin of the right conal artery (yellow arrow). MAc: marginal acute; RCA: right coronary artery.

Although usually two coronary arteries (right and left) are seen emerging from the aorta, three or even four independent origins have been described. In these cases, frequently (36% of individuals) the right conal artery is the one with an independent origin² (Figure 2.4). Also, it is not rare to find separate origins for both main branches of the left artery, this implying the absence of left main trunk⁴. On the other hand, the origin of both arteries from a single coronary sinus has been described, either from a single *ostium* or from separate orifices at the same sinus².

2.2 Left Coronary Artery

The left coronary artery is a large artery (Figure 2.5) with an approximate diameter of

5 mm at its origin, that supplies an extensive portion of the walls of the left chambers of the heart, with blood including most of the interventricular septal mass².

2.2.1 Left main (LM) artery

A common initial segment of the left coronary artery, the LM has variable length (Figure 2.6), is embedded in adipose tissue, and courses between the main pulmonary artery and the left atrial appendage^{3,5} (Figures 2.7–2.8). Rarely (<1% of individuals), the LM is absent (Figure 2.9), with independent origins of its main branches from the left coronary sinus⁴.

In addition to its main ramification, the LM emits no other branches, except in those rare instances where the artery of the sinus node originates from it². At the level of the left atrio-ventricular groove, the LM gives two or three

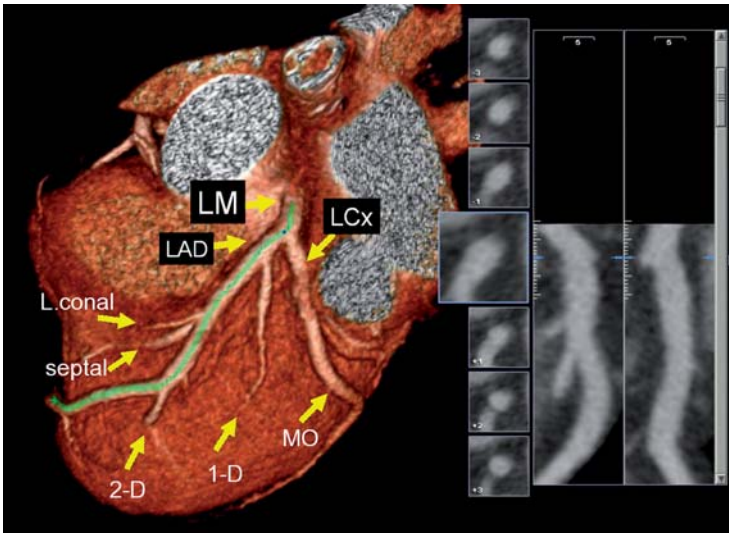


FIGURE 2.5 Left coronary artery with its branches, as seen on a 3D (left) and a multiplanar reconstruction (MPR) of its proximal segment (right). LAD: left anterior descending; L.Conal: left conal branch; LCx: left circumflex; LM: left main; MO: marginal obtuse branch of the LCx; Septal: first septal anterior branch of the LAD; 1D and 2D: first and second diagonal branches of the LAD.

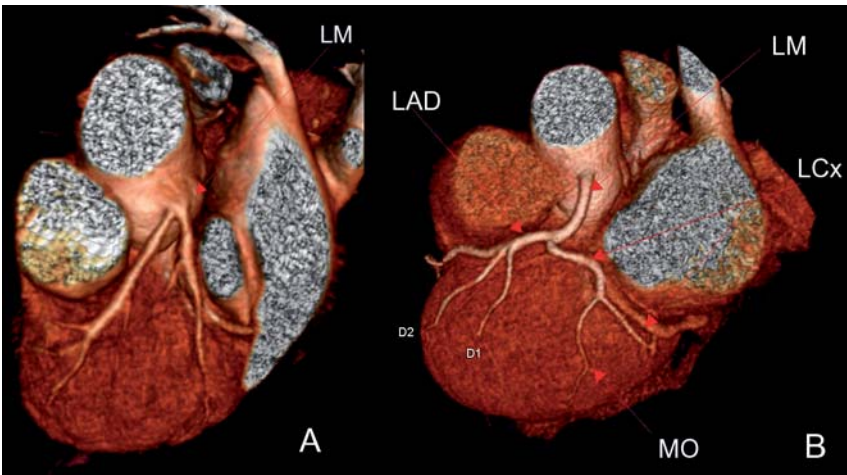


FIGURE 2.6 Short (A) and long (B) normal variants of left main (LM) coronary artery. LAD: left anterior descending; LCx: left circumflex; MO: marginal obtuse branch of the LCx; 1D and 2D: first and second diagonal branches of the LAD.

branches, namely the left anterior descending, the left circumflex artery and, occasionally, the intermediate artery³.

2.2.2 Left anterior descending (LAD)

LAD coronary artery is a large vessel—4–5 mm in diameter at its proximal portion—that occupies the anterior interventricular groove, running in parallel with the great cardiac vein

(Figure 2.10), with which it exhibits crossover points^{2,6}. It usually extends to the apical region of the left ventricle and, in two thirds of individuals, it reaches the distal (Figure 2.11) or even the middle portion of the posterior interventricular groove⁷. In these cases, the LAD frequently shows anastomotic connections with the left posterior descending artery (PDA)².

The LAD gives some branches along its course^{2–4,6,8–12}:

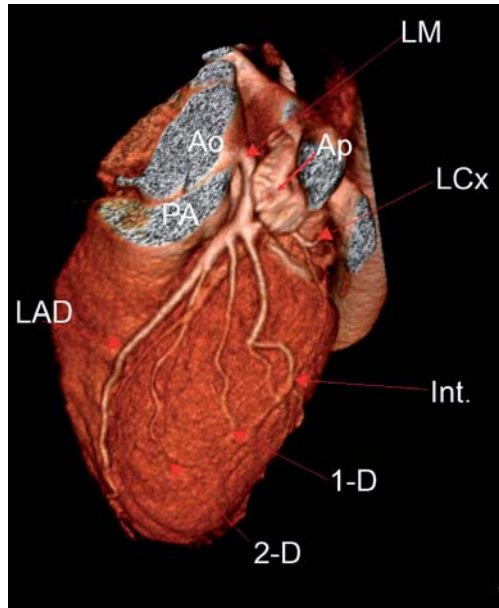


FIGURE 2.7 Anatomical relationships of left main (LM) coronary artery. Ao: aorta; Ap: left atrial appendage; Int.: intermediate artery; LAD: left anterior descending; LCx: circumflex artery; PA: pulmonary artery; 1D and 2D: first and second diagonal branches of the LAD.

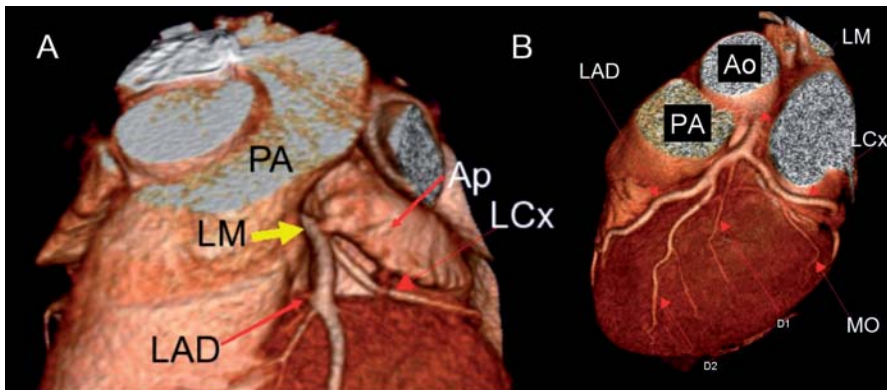


FIGURE 2.8 Anatomical relationships of the proximal segments of the main branches of the left coronary artery. Observe the course of the vessels below the left atrial appendage (Ap) in A, which is adequately displayed when the Ap is removed, in B. Ao: aorta; LAD: left anterior descending; LCx: circumflex artery; LM: left main; MO: marginal obtuse branch of the LCx; PA: pulmonary artery; 1D and 2D: first and second diagonal branches of the LAD.

- *Left conal artery* (Figure 2.5): with an origin in the proximal LAD, it communicates with the right conal artery, with which it constitutes the “arterial ring of Vieussens,” along with the *vasa vasorum* of the aorta and pulmonary artery.
- *Right anterior ventricular branches*: usually irrelevant in number and diameter, as the right ventricle is almost exclusively irrigated through the right coronary artery.
- *Left anterior ventricular branches (diagonal arteries)*^{13–15} (Figure 2.7): Variable in number, these branches distribute diagonally over the anterior aspect of the left ventricle (Figure 2.12). The origin of the first diagonal artery (Figure 2.5) is used as the anatomical point dividing the middle and distal segments of the LAD. Frequently, one of these diagonal arteries is particularly large and follows a course parallel to the

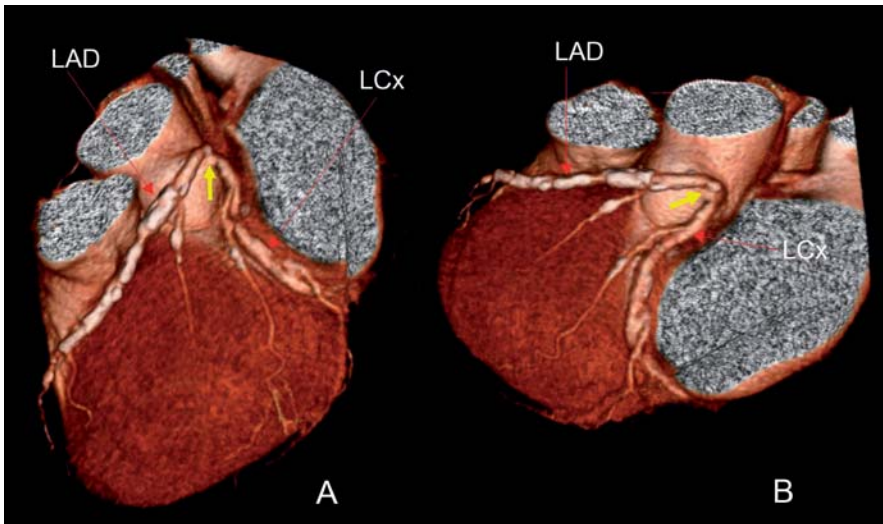


FIGURE 2.9 Left oblique (A) and left lateral (B) views in a case of absent left main (LM) (yellow arrow), with an independent origin of the left anterior descending (LAD) and circumflex (LCx) arteries. Observe the extensive vessel wall calcification.

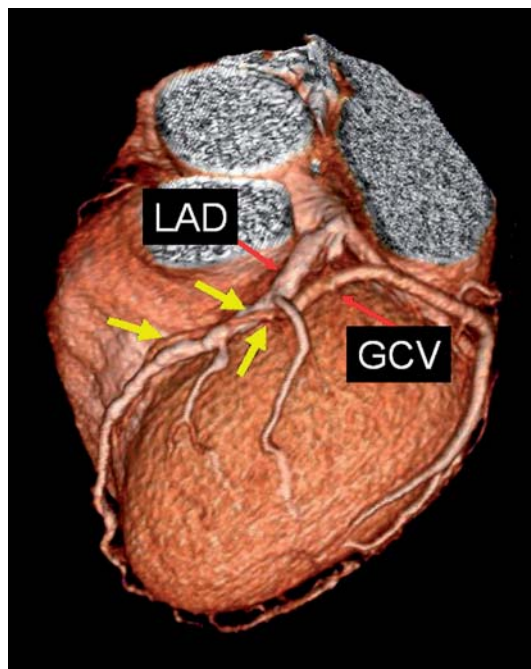


FIGURE 2.10 Relationship between the left anterior descending (LAD) and the great cardiac vein (GCV), showing vessel crossing at their middle course (yellow arrows).

LAD (Figure 2.12D), from which it can be distinguished by the lack of septal branches and the presence of secondary small diagonal branches. In cases where this diagonal artery reaches the obtuse margin of the heart, and from there, the posterior aspect of the left ventricle, it is known as posterolateral

artery. The absence of diagonal arteries is extremely rare and, thus, when this is the case in coronary angiography, occlusion of some of these branches can be suspected.

- *Anterior septal branches*: variable in number, these branches arise orthogonally from the LAD and distribute into the anterior two-

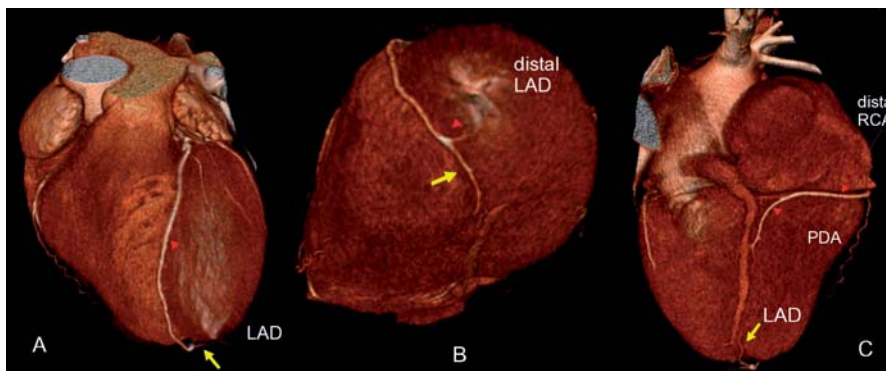


FIGURE 2.11 Recurrent course of the distal segment of the left anterior descending (LAD) artery reaching the interventricular posterior groove. A: anterior view; B: apical view; C: posterior view. PDA: posterior descending artery.

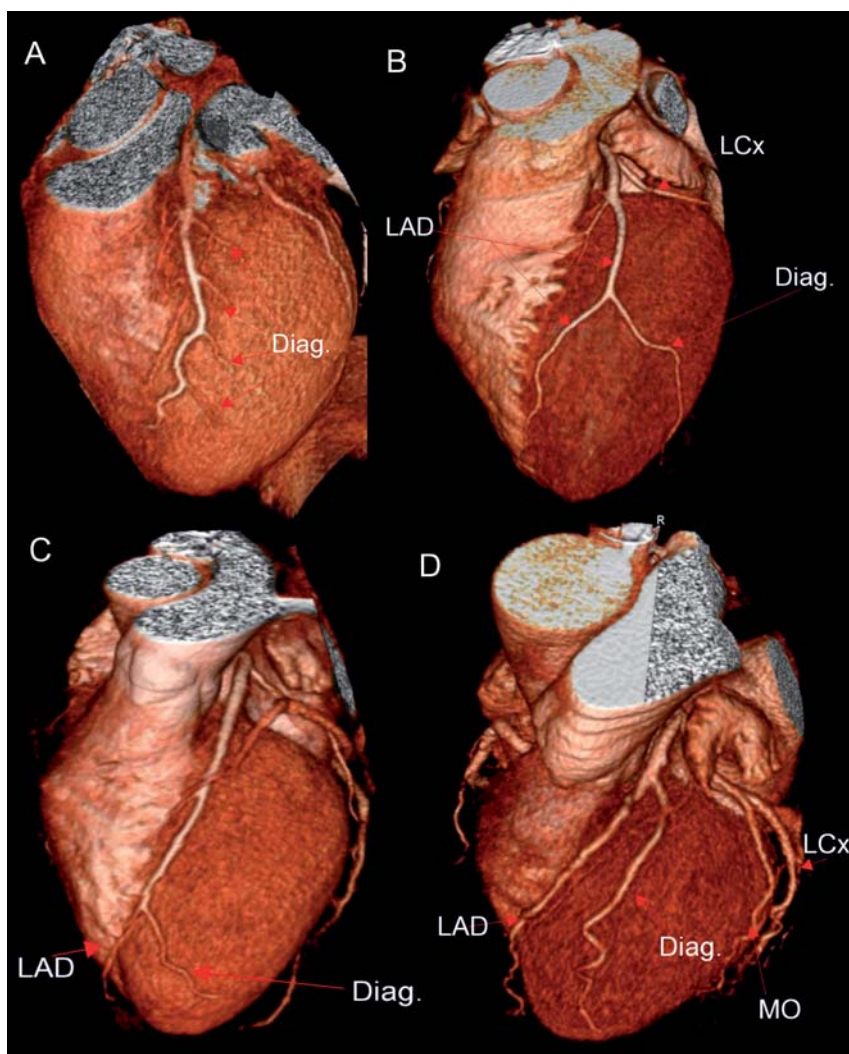


FIGURE 2.12 Normal anatomical variants of diagonal (Diag) branches. A: Multiple small brief branches; B: Single branch emerging from the middle left anterior descending (LAD); C: Single branch emerging from the distal LAD; D: Large vessel coursing parallel to the LAD; LCx: left circumflex; MO: marginal obtuse branch.

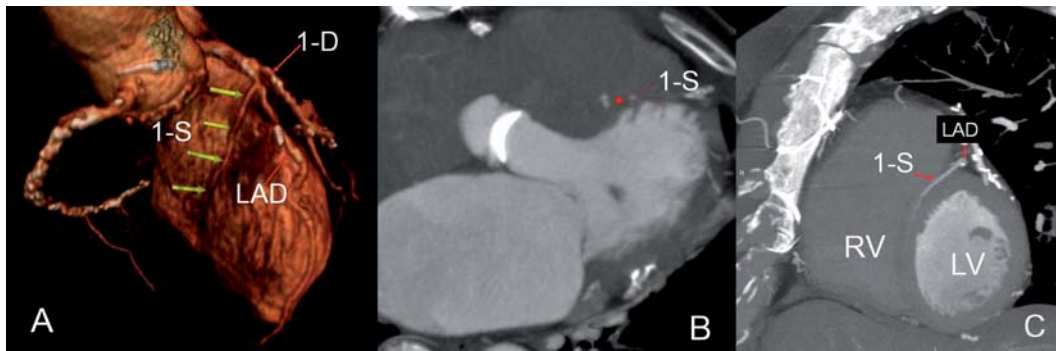


FIGURE 2.13 First septal branch (1-S) of the left anterior descending (LAD). A: The weak contrast opacification of the right heart chambers allows the visualization of the course of 1-S through the interventricular septum; B and C: Maximal Intensity Projection (MIP) images showing transverse (B) and longitudinal (C) sections of 1-S into the septum; LV: left ventricle; RV: right ventricle.

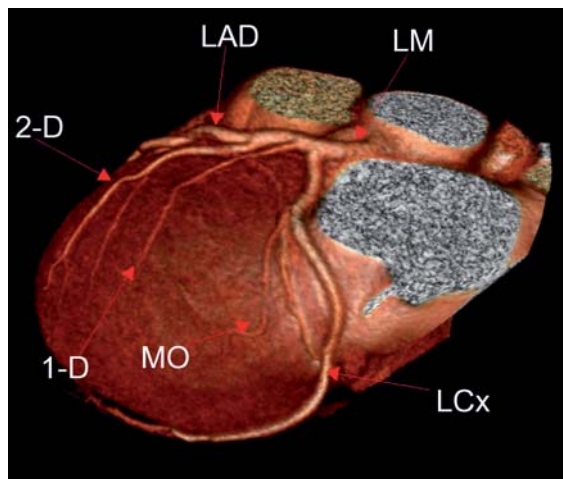


FIGURE 2.14 Left circumflex (LCx) artery ending at the (left) obtuse margin of the heart. LAD: left anterior descending; LM: left main; MO: marginal obtuse; 1D and 2D: first and second diagonal branches.

thirds of the interventricular septum. The first septal branch is usually a well developed vessel (Figure 2.13), its origin being considered as the reference point dividing the proximal and middle portions of the LAD. Rarely, this first septal branch courses closely parallel to the LAD.

2.2.3 Left circumflex (LCx)

LCx artery is also a large vessel, similar in diameter to the LAD, although more variable in terms of length and anatomical distribution^{3,4}. The proximal portion of the vessel lies beneath the left atrial appendage and, from there, its course follows the anterior aspect of the left atrioventricular groove, ending at the obtuse

margin of the heart (Figure 2.14). In some cases, the vessel extends to the posterior aspect of the left atrioventricular groove, usually below the coronary venous sinus, ending proximally to the region of the *crux cordis*²⁻⁴. Finally, in cases of anatomical dominance of the left coronary system, the LCx goes beyond this region and gives the PDA.

The LCx gives origin to different branches during its course^{2-4,6,8-12}:

- *Anterior or anterolateral ventricular branches*: when present, these small vessels (Figures 2.15, 2.16A) arise proximally and course parallel to the first diagonal artery. When this artery is absent, it is substituted by these branches.

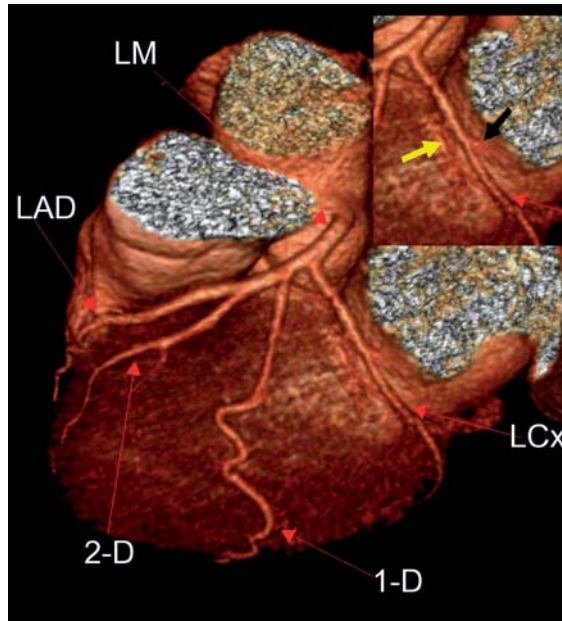


FIGURE 2.15 Left circumflex (LCx) artery, with a small anterolateral branch (yellow arrow) and an atrial branch (black arrow) (see inset at top right). LAD: left anterior descending; LM: left main; 1D and 2D: first and second diagonal branches.

- *Sinusal or sinoatrial branch*^{16,17} (Figure 2.16): although usually arising from the right artery, the sinusal branch emerges from the proximal segment of the LCx in 30–35% of individuals, courses around the left atrium, and reaches the sinus node region at the superior vena cava drainage.
- *Atrial arteries* (Figure 2.15): these small vessels are usually located beneath the base of the left atrial appendage or at the posterior aspect of the left atrium.
- *Obtuse marginal branches* (Figure 2.17): usually one or two, their origin is used as a reference dividing the proximal and medial segments of the LCx. These branches are well-developed vessels emerging orthogonally from the LCx and coursing along the left margin of the heart until they reach the apex, where they can communicate with vessels from the LAD.
- *Posterior ventricular branches* (Figures 2.17A, 2.18, 2.19): although the posterior wall of the left ventricle is mostly irrigated by branches from the right PDA, when this vessel is absent, a variable number of these posterior ventricular branches—together with a number of interventricular

branches of the LCx—are responsible for the blood supply to this region.

- *Atrioventricular nodal branch*: it arises from the LCx in up to 20% of subjects, particularly in cases of left dominance.

2.2.4 Intermediate coronary artery

In a proportion of individuals, reported as between 25–40%, the LM divides into three branches; in addition to the LAD and the LCx, a third vessel is found, known as median or intermediate artery, arising from the vertex of the angle formed by the two former arteries⁴ (Figure 2.20).

Usually a large vessel, the intermediate artery runs over the antero-lateral aspect of the left ventricle, giving septal anterior branches (Figure 2.21) as well as to the anterior papillary muscle. The length of the vessel is variable, although frequently it ends near the obtuse left margin of the heart. Not rarely, however, it reaches the apex or even the inferior aspect of the left ventricle.

In those cases with a largely developed intermediate artery, the diagonal and obtuse marginal arteries are, accordingly, smaller vessels.

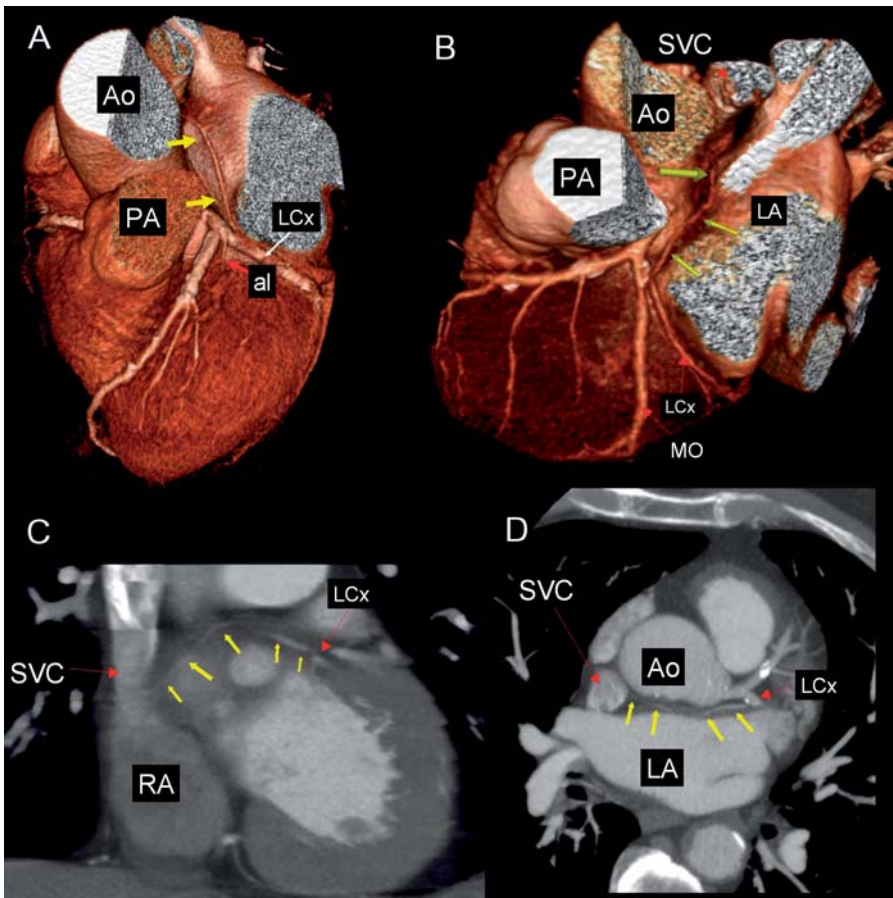


FIGURE 2.16 Left circumflex (LCx) artery. A: Anterior view showing an anterolateral (al) branch (red arrow) and a sinus branch (yellow arrows); B: Cranial view also displaying the sinus branch (yellow arrows); C: MPR on an oblique view with volume render, and; D: Axial slice with MIP, both showing the sinus branch of the LCx and its course towards the region of the superior vena cava (SVC); Ao: aorta; LA: left atrium; MO: marginal obtuse branch; PA: pulmonary artery; RA: right atrium.

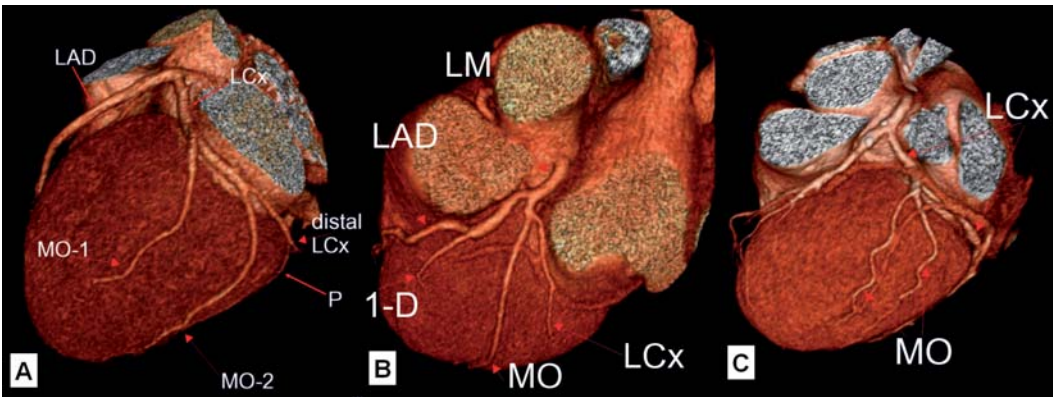


FIGURE 2.17 Anatomy of marginal obtuse (MO) branches. A: Two MO branches are seen (1 and 2) and, also, a posterior branch irrigating the posterior aspect of the left ventricle; B: Occasionally, only a single MO branch is present which arises early from the left circumflex (LCx) and is frequently larger than the LCx itself; C: Bifurcated MO branch; LAD: left anterior descending; LM: left main; 1-D: first diagonal.

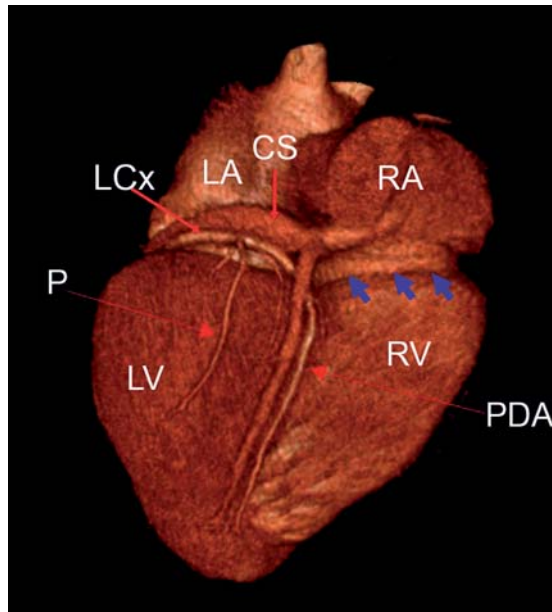


FIGURE 2.18 Anatomical dominance of the left coronary system, with a posterior descending artery (PDA) depending from the left circumflex (LCx); a posterior (P) branch of the LCx is seen irrigating the posterior aspect of the left ventricle (LV), while the right coronary artery does not reach the inferior aspect of the heart (blue arrows). CS: coronary sinus; LA: left atrium; RA: right atrium; RV: right ventricle.

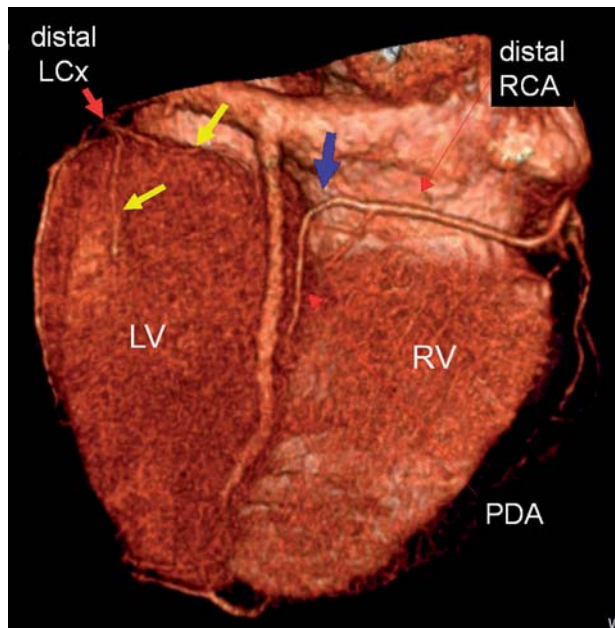


FIGURE 2.19 Anatomical dominance of the right coronary system where the right coronary artery (RCA) gives origin to the posterior descending artery (PDA) but not to posterolateral branches (blue arrow). This myocardial territory depends, in this case, from posterior branches (yellow arrows) emerging from the distal left circumflex (LCx). LV: left ventricle; RV: right ventricle.

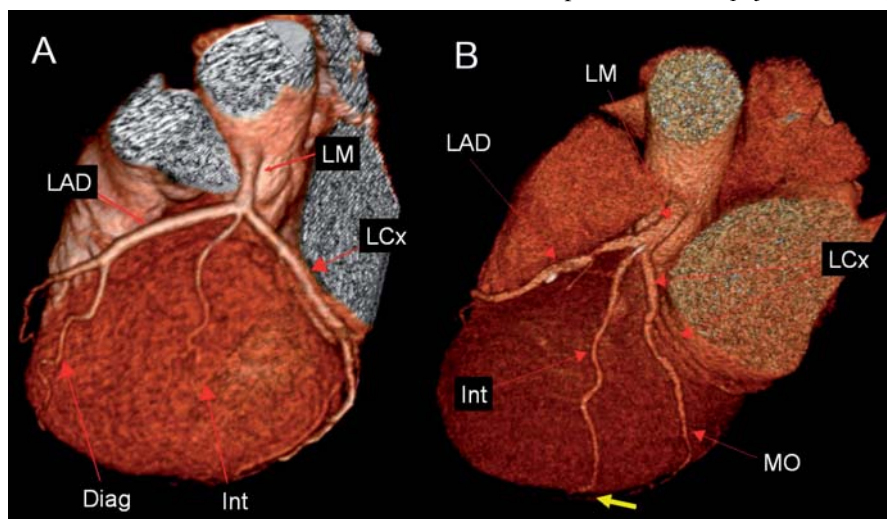


FIGURE 2.20 Intermediate (Int) coronary arteries from two different subjects: in case B, the vessel is large, reaching the left margin of the heart (yellow arrow). Diag: diagonal branch; LAD: left anterior descending; LCx: circumflex artery; LM: left main; MO: marginal obtuse branch.

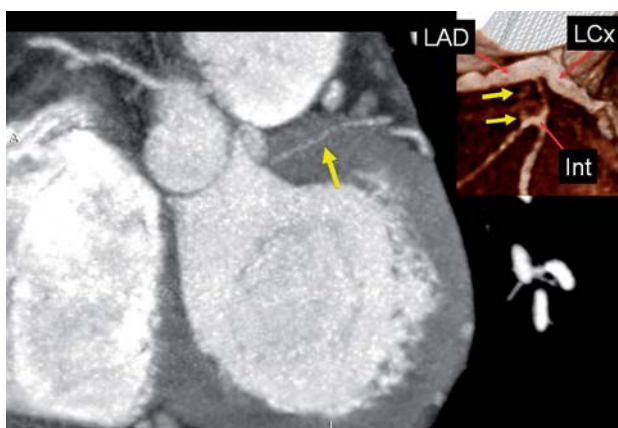


FIGURE 2.21 Example of septal branch originated from one of the bifurcating branches of an intermediate (Int) artery (yellow arrows, at the inset, top right) with an intramyocardial course through the anteroseptal wall (arrow on the large panel). LAD: left anterior descending; LCx: circumflex artery.

2.3 Right Coronary Artery (RCA)

The RCA supplies the blood flow for the right atria and ventricle and, when dominant, also for a variable extension of the posterior aspect of the left ventricle.

Originated in the right coronary sinus, the proximal segment of the RCA courses closely to the right atrial appendage and is then located on the anterior aspect of the right atrioventricular groove, where it is embedded in adipose tissue (Figure 2.22). At its medial segment, the RCA

rounds the right acute margin of the heart and through the posterior aspect of the right atrioventricular groove, it reaches the region of the *crux cordis*^{2-4,6,8} (Figure 2.23).

There are variants of this anatomical distribution: in 10% of individuals the RCA ends at the level of the acute margin of the heart (Figure 2.24), or between this region and the *crux cordis*; in 60% the RCA extends beyond the *crux cordis* and reaches the inferior wall of the left ventricle (Figure 2.23A), where it shows connections with the distal LCx artery; finally, in 20% of subjects the vessel arrives

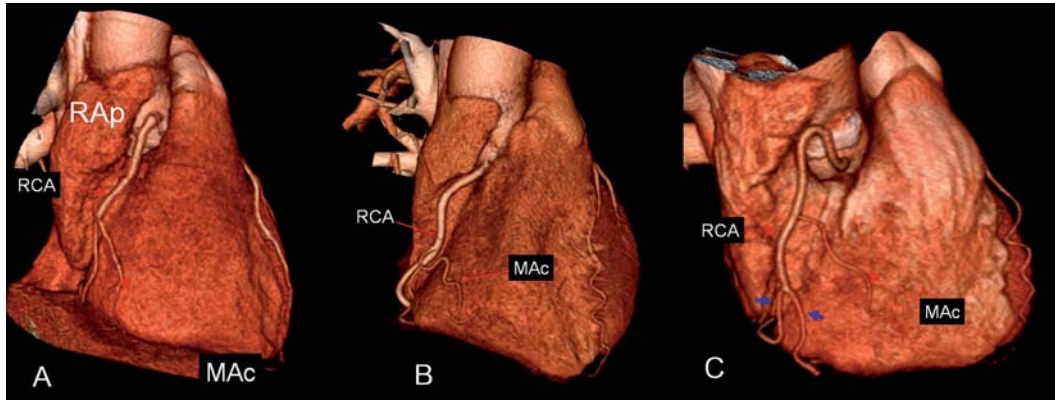


FIGURE 2.22 Anatomy of the right coronary artery (RCA). A: Proximal and middle segments of the vessel coursing in close relationship with the right atrial appendage (RAp) and giving origin to the marginal acute branch (MAc); B: Example of a tortuous MAc; C: Early bifurcation of the RCA at its middle segment (blue arrows).

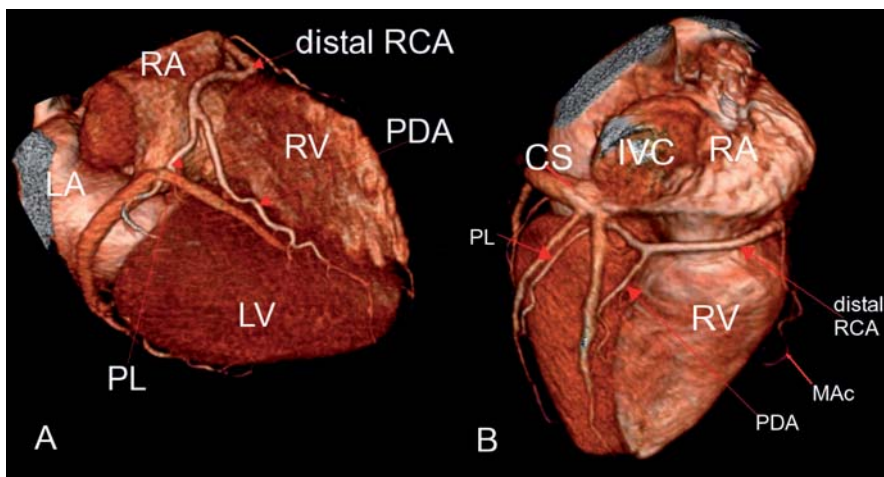


FIGURE 2.23 Anatomy of the distal right coronary artery (RCA). A: Bifurcation of the vessel near the region of the *crux cordis* into a posterior descending artery (PDA) and a posterolateral (PL) branch; B: Example of a long PL branch reaching the left margin of the heart; a marginal acute (MAc) artery is also seen over the right margin; CS: coronary sinus; IVC: inferior vena cava; LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

to the left cardiac margin, irrigating the area corresponding to the LCx (Figure 2.23B).

The RCA gives different branches along its course^{2-4,6,8-12}:

- *Right conal branch*: as mentioned earlier, in up to 36% of individuals, this vessel shows an origin in the anterior aspect of the right coronary sinus, independent from the one of the RCA (Figure 2.4). Usually a small vessel, the right conal branch may occasionally be large, supplying an extense portion of the right ventricle, in which case, the RCA appears less developed (Figure 2.25). As described above, the right and left conal

branches are connected, constituting the “arterial ring of Vieussens.”

- *Sinus node branch*^{16,17} (Figure 2.26): this vessel originates from the RCA in more than 50% of individuals, usually arising from the most proximal portion of the RCA or, rarely, from its middle or even distal segment. The sinus node branch courses over the base of the right atrial appendage, ending at the drainage of the superior vena cava into the right atrium.
- *Atrial branches*: variable in number and size, these branches are distributed over the anterolateral aspect of the right atrium, although a posterior branch also does exist,

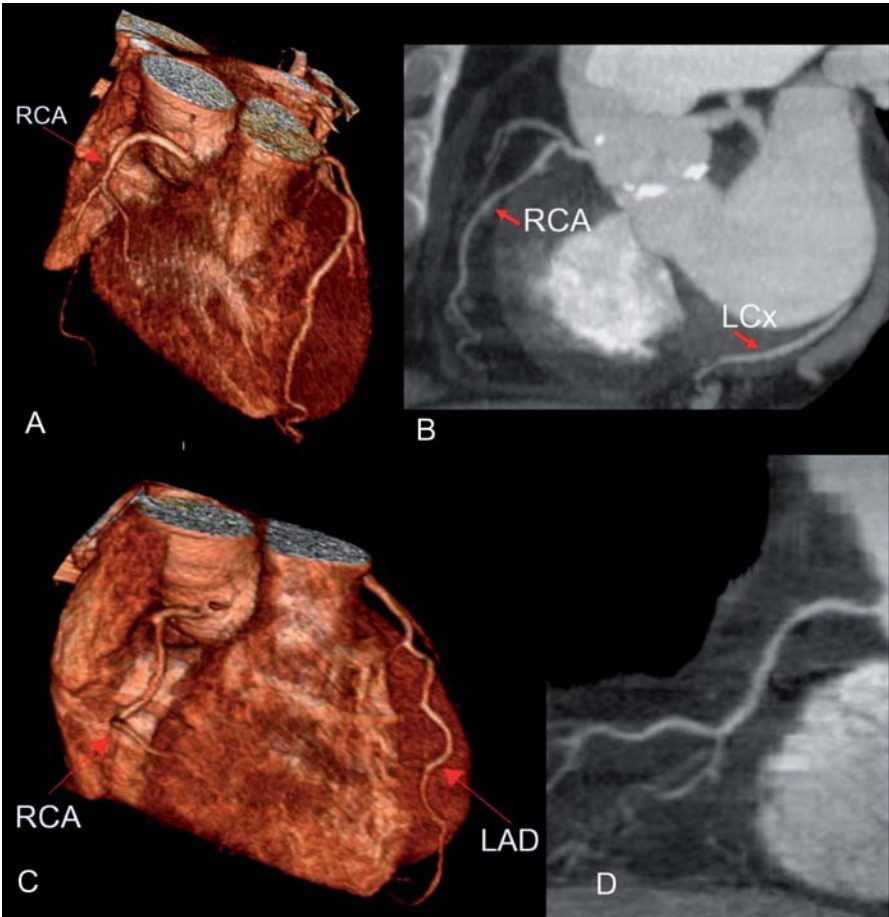


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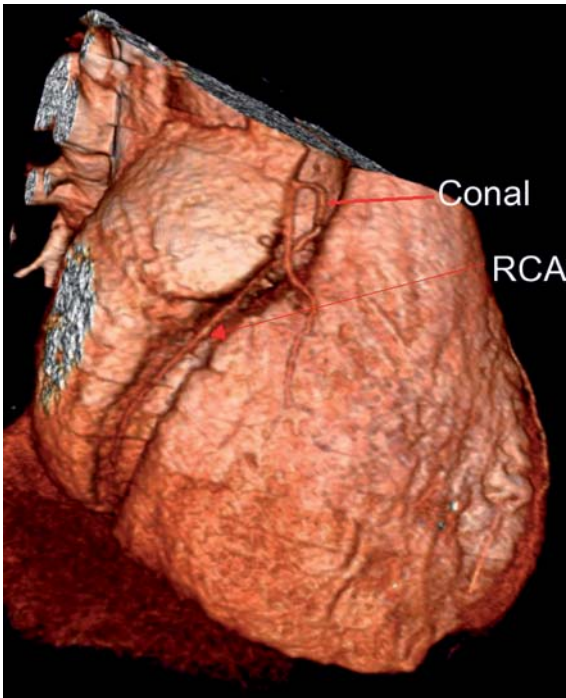


FIGURE 2.25

- F. 2.24. Anatomical dominance of the left coronary system with poorly developed right coronary artery (RCA), not reaching the right margin of the heart. Two cases are presented (A-B and C-D) with 3D volume rendering images (left panels) and curved MPR (right panels). LAD: left anterior descending; LCx: circumflex artery.
- F. 2.25. Right conal artery in a case with a relatively narrow right coronary artery (RCA).

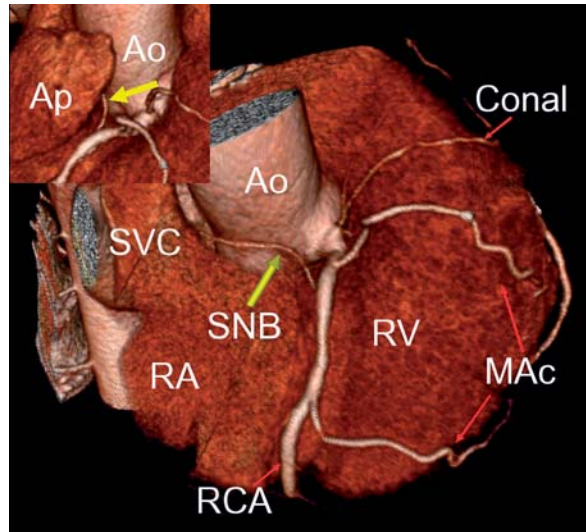


FIGURE 2.26 Sinus node branch (SNB) coursing close to the right atrial appendage (Ap) (see inset, at top left) and ending near the region of drainage of the superior vena cava (SVC). Ao: aorta; MAC: marginal acute branch; RA: right atrium; RV: right ventricle.

- supplying both atria or the posterior left atrium exclusively.
- *Acute marginal branch*: this anterior right ventricular branch is usually a well-developed vessel, coursing over the right ventricular free wall (Figure 2.27), near the right acute margin of the heart, and reaching the region of the apex in most individuals. Not infrequently, one or two additional marginal branches are seen emerging from the RCA and running parallel to the acute marginal branch, which is, however, the larger vessel (Figure 2.28).
 - *Posterior right ventricular branches*: these are small vessels—not always present—arising from the distal RCA, and irrigating the inferior aspect of the right ventricle (Figure 2.29). Their degree of development is inverse to that of the acute marginal branch, which occasionally distributes over the same region (Figure 2.30).
 - *Interventricular posterior branch (or right PDA)*: this artery is a branch of the RCA in up to 90% of individuals, arising from the *crux cordis*, or the region where both

posterior atrioventricular grooves meet with the posterior interventricular groove. In nearly 70% of subjects, the right PDA is a single branch (Figure 2.23A) coursing along the posterior interventricular groove, ending next to the most distal recurrent branch of the LAD, in the region of the apex. In the remaining 30% of cases, 2 or 3 smaller branches are present, coursing in parallel at both sides of the interventricular groove (Figure 2.31). In some cases, the right PDA originates from the RCA at some point between the right acute cardiac margin and the *crux cordis*, coursing diagonally over the inferior aspect of the right ventricle (Figure 2.32). The PDA irrigates the posterior aspect of both ventricles, also giving small posterior septal branches in those cases when the vessel courses along the posterior interventricular groove. These branches penetrate into the posterior septum in a lesser extension than the anterior ones and are usually lacking at the apical region. The first of these posterior septal branches is frequently a well-developed vessel emerging

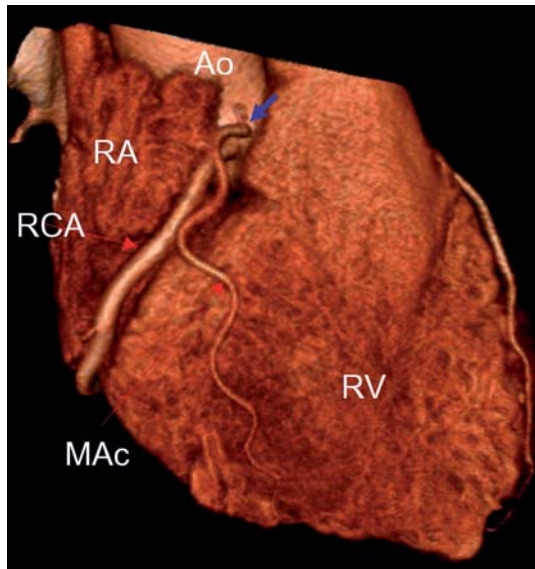


FIGURE 2.27 Independent origin of a marginal acute (MAC) branch from the aorta (Ao) (blue arrow). RA: right atrium; RCA: right coronary artery; RV: right ventricle.

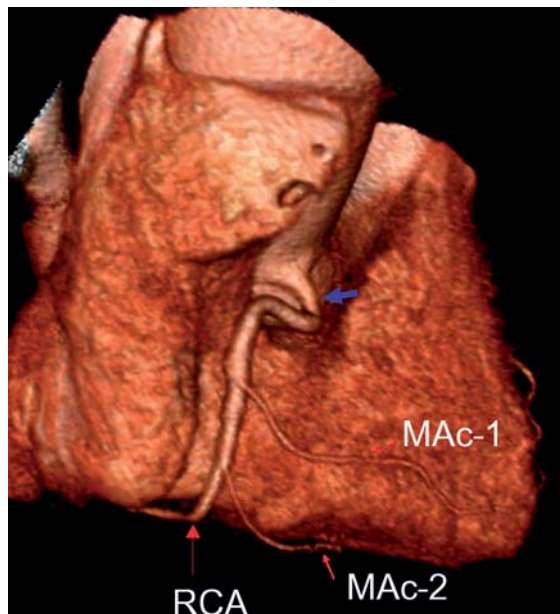


FIGURE 2.28 Marginal acute (MAC-1, MAC-2) branches of the right coronary artery (RCA), which shows a tortuous course at its proximal segment (blue arrow). Observe the long course of the MAC-1, reaching the cardiac apex.

at the level of the *crux cordis*, irrigating the atrioventricular node (Figure 2.33).

- *Right posterobasal or posterolateral arteries:* widely variable in number, size, and distribution pattern, these vessels usually course over the inferior aspect of the left ventricle (Figure 2.34).

2.4 Pattern of Dominance of the Coronary Arteries

The term “dominant” is widely used in the clinical setting as referred to the coronary artery that reaches the *crux cordis* and gives origin to

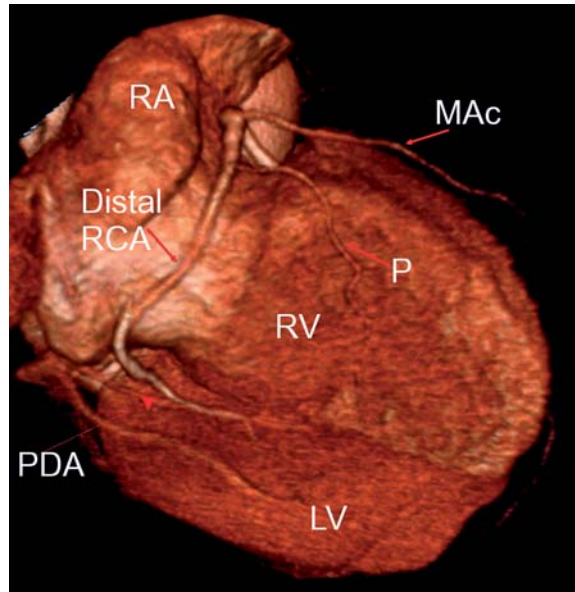


FIGURE 2.29 Posterior right ventricular branch (P). MAc: marginal acute branch; LV: left ventricle; PDA: posterior descending artery; RA: right atrium; RCA: right coronary artery; RV: right ventricle.

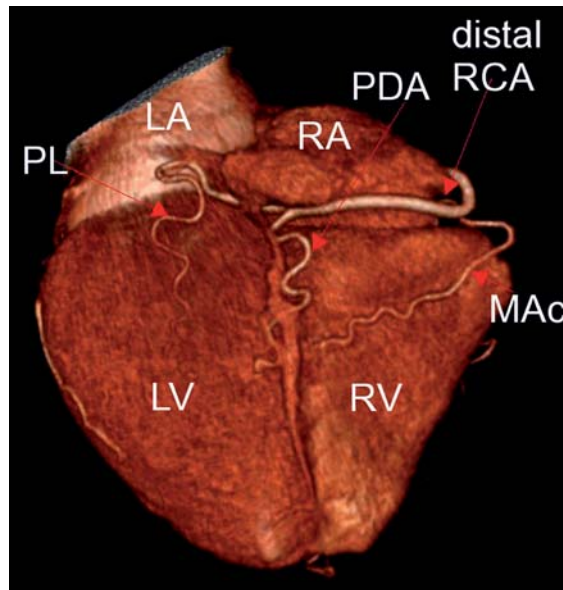


FIGURE 2.30 Marginal acute (MAc) branch of the right coronary artery (RCA) extending into the inferior aspect of the right ventricle similarly to the posterior right ventricular branch (see Figure 2.29). LA: left atrium; LV: left ventricle; PDA: posterior descending artery; PL: posterolateral branch; RA: right atrium; RV: right ventricle.

the PDA. However, this expression can falsely lead to the assumption that a particular coronary artery is responsible for the irrigation of most of the ventricular myocardial mass, while it is the left coronary artery (whether or not

“dominant”), which actually supplies the largest amount of ventricular myocardium in most normal hearts^{2-4,18}.

Anatomical macroscopic exams show a relatively constant pattern of distribution of the

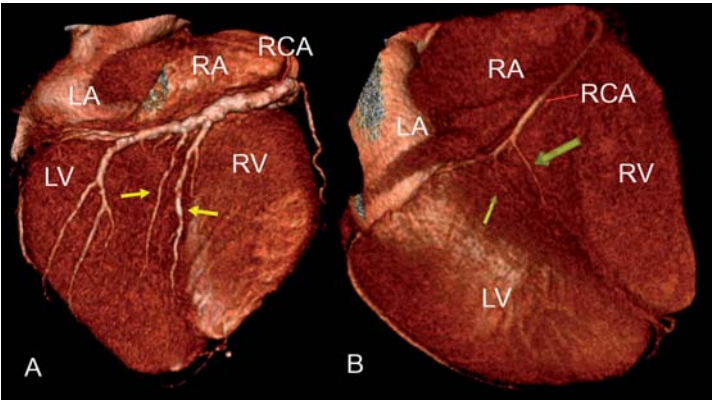


FIGURE 2.31 Examples of posterior descending arteries (PDA): sometimes not a single vessel, but consisting of two or more branches (arrows) of large (A) or small (B) diameter, arising from the distal right coronary artery (RCA). LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

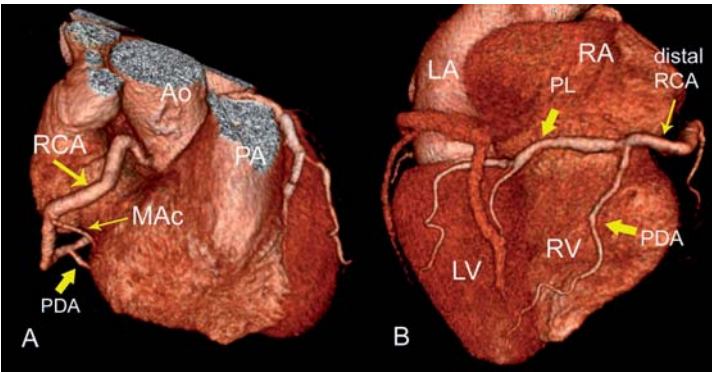


FIGURE 2.32 Anterior (A) and posterior (B) views from a subject with a posterior descending artery (PDA) originating from the right coronary artery (RCA) proximally to the region of the *crux cordis*, presenting with a diagonal course over the inferior wall of the right ventricle. Ao: aorta; MAC: marginal acute branch; LA: left atrium; LV: left ventricle; PA: pulmonary artery; RA: right atrium; RV: right ventricle.

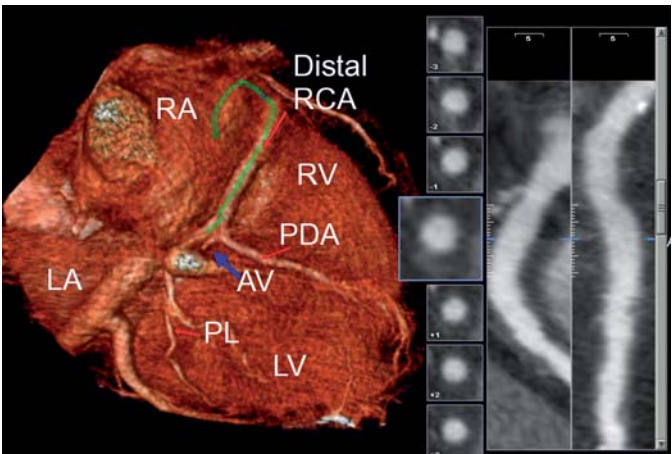


FIGURE 2.33 Atrioventricular (AV) node artery (blue arrow) originating from the right coronary artery (RCA) at the level of the *crux cordis*. LA: left atrium; LV: left ventricle; PDA: posterior descending artery; PL: posterolateral branch; RA: right atrium; RV: right ventricle.

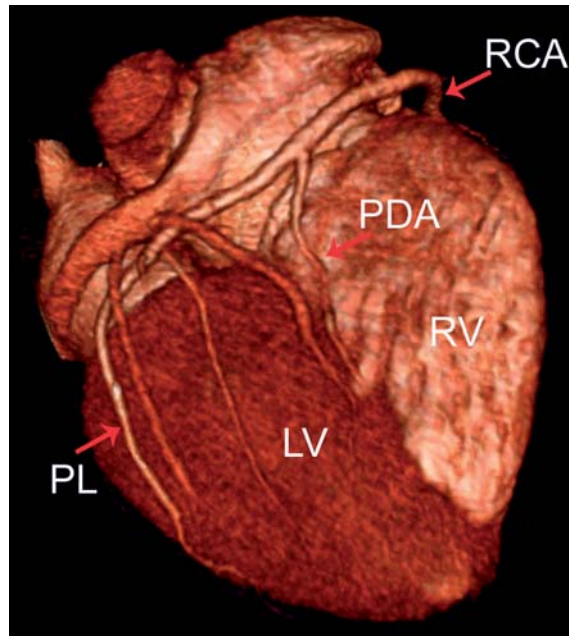


FIGURE 2.34 Large posterolateral branch (PL) from the right coronary artery (RCA) reaching the left margin of the heart. LV: left ventricle; PDA: posterior descending artery; RV: right ventricle.

coronary circulation at the level of the anterior aspect of the heart, in contrast with the inferior aspect, where there is more variability, as blood supply is provided by the distal RCA and LCx, both with a degree of development which is complementary to each other.

Three patterns of anatomical coronary artery dominance can be distinguished (Baroldi and Scomazzoni, 1965):

- *Type I* (77% of individuals): in this case, the RCA gives origin to the PDA, and in turn, may end up with different patterns: (a) at the posterior interventricular groove, rarely with branching vessels for the left ventricle (5%) (Figure 2.19); (b) providing distinctive medium-sized branches for a portion of the inferior aspect of the left ventricle (55%) (Figure 2.23A); and (c) with large branches for most of the inferior aspect of the left ventricle, reaching the left cardiac margin (17%) (Figure 2.23B).
- *Type II* (8%): in this case, the PDA is a branch of the LCx, appearing either as a single or a ramified vessel (Figures 2.18, 2.35).
- *Type III* (“balanced” circulation) (15%): in this case there are two PDA, one provided for each vessel (RCA and LCx), coursing in parallel to the posterior interventricular groove (Figure 2.36). In this pattern, the

apex of the heart and the inferior aspect of the interventricular septum are irrigated by the left coronary artery.

A knowledge of these patterns of anatomic dominance is fundamental for an appropriate assessment of a coronary angiography, where a relatively frequent cause of concern is the distinction between a branch occlusion and a normal anatomical pattern of the PDA.

2.5 Congenital Anomalies of the Coronary Arteries

Congenital anomalies of the coronary arteries comprise a number of entities where features different from those considered as defining normalcy are present¹⁹ (Table 2.1).

Despite the large number of reported anomalies^{20–22} (Table 2.2), congenital anomalies of coronary arteries are present with low prevalence rates; lower than 3% of all congenital heart diseases, and less than 1% of the general population²³. Congenital anomalies of coronary arteries may be present in the absence of other structural heart disease^{24–26} or, more frequently, they are found in patients with D-transposition of the great vessels^{27–29}

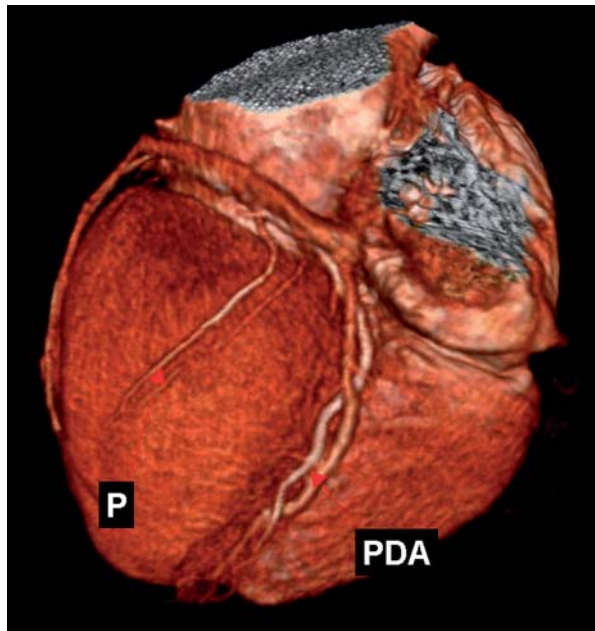


FIGURE 2.35 Posterior descending artery (PDA) originating from the circumflex artery in a subject with “left dominance.” P: posterior ventricular branch.

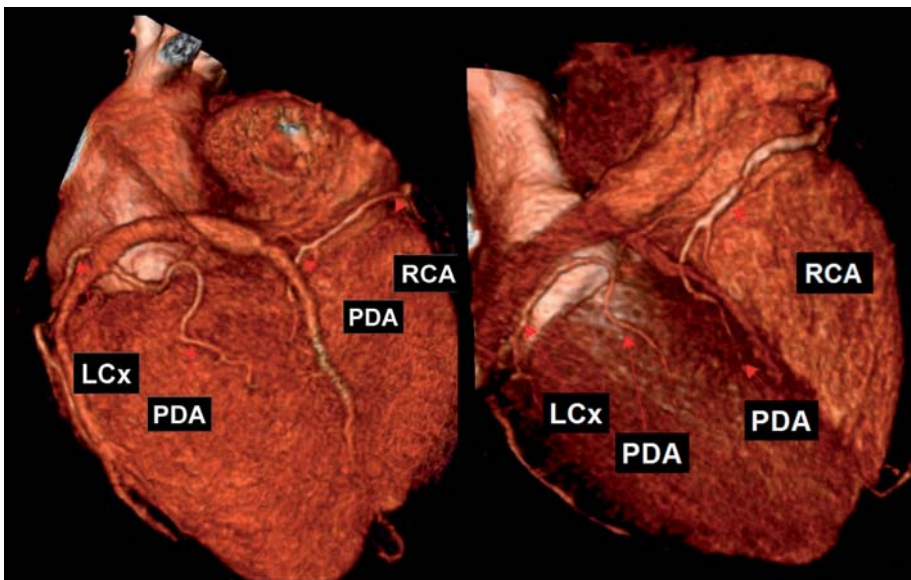


FIGURE 2.36 Two different subjects with a balanced coronary circulation pattern: both the right coronary artery (RCA) and the left circumflex artery (LCx) give origin to a posterior descending artery (PDA) from their own.

(Figure 2.37), Fallot’s tetralogy, pulmonary atresia with ventricular septal defect, or, in general, in patients with chromosomal abnormalities (trisomy 18)^{28,30}. Not rarely, they are incidental findings in coronary angiographies performed due to symptoms not related with these abnormalities.

Congenital anomalies of the coronary arteries may be the cause of myocardial ischemia and even sudden death³¹. For this reason, it is clinically important to recognize two different types of these anomalies³², depending on whether or not they are able to produce myocardial ischemia.

Table 2.1 Normal coronary artery anatomical features and corresponding congenital anomalies (adapted from Trivellato¹⁹).

	Normal	Abnormal
• Coronary <i>ostia</i>	Two	One
• Course of main trunks	Epicardial	Intramural
• Course of RCA	Right AV groove	Left side (partially)
• Course of LCA	Left AV and interventricular anterior grooves	Right side (partially)
• PDA origin	RCA or LCx	LAD
• Coronary artery drainage	Capillary bed	Other

Table 2.2 Congenital coronary artery abnormalities (adapted from Angelini^{20–22}).

- A. Anomalies of the origin and course of vessels
 1. Absent left main trunk
 2. Anomalous location of a coronary *ostium* within the proper coronary sinus
 3. Anomalous location of a coronary *ostium* at places other than the normal coronary sinuses
 - a. Right posterior aortic sinus
 - b. Thoracic aorta, supraortic vessels, and their branches
 - c. Left and right ventricle
 - d. Pulmonary artery (Bland-White-Garland syndrome, in case of LCA)
 4. RCA/LCA arising from the contrary sinus, with anomalous course:
 - a. Posterior atrioventricular groove or retrocardiac
 - b. Retroaortic
 - c. Between aorta and pulmonary artery
 - d. Intraseptal
 - e. Anterior to pulmonary outflow or precardiac
 - f. Posteroanterior interventricular groove
 5. Single coronary artery (40% associated to other congenital heart diseases)
- B. Anomalies of intrinsic coronary arterial anatomy
 1. Congenital ostial stenosis or atresia
 2. Absent coronary artery
 3. Coronary hypoplasia
 4. Intramural coronary artery (muscular bridge)
 5. Subendocardial coronary course
 6. Coronary crossing
 7. Anomalous origin of PDA from a branch of the LAD or from a septal penetrating branch
 8. Absent LAD or PDA
 9. Ectopic origin of first septal branch
- C. Anomalies of coronary drainage
 1. Inadequate arteriolar/capillary ramifications
 2. Fistulas from RCA or LCA
- D. Anomalous collateral vessels

2.5.1 Anomalies relatable to myocardial ischemia

In most of these anomalies, a segment of a coronary artery is seen to course abnormally between the wall of the aortic root and that of the main pulmonary artery^{33–36} (Figures 2.38–2.40). This situation may cause exertional myocardial ischemia due to enlargement of the great vessels, secondary to transiently increased flow³². Also, in cases of abnormal coronary

artery course around the anterior aspect of the pulmonary artery (Figure 2.41), ischemia may be present in those situations of enlargement of the right ventricular outflow tract, as in pulmonary hypertension^{37,38}.

Abnormal origin of coronary arteries from the pulmonary artery, either the LCA (Bland-White-Garland syndrome)^{39,40} or the RCA, is also an obvious cause of myocardial ischemia.

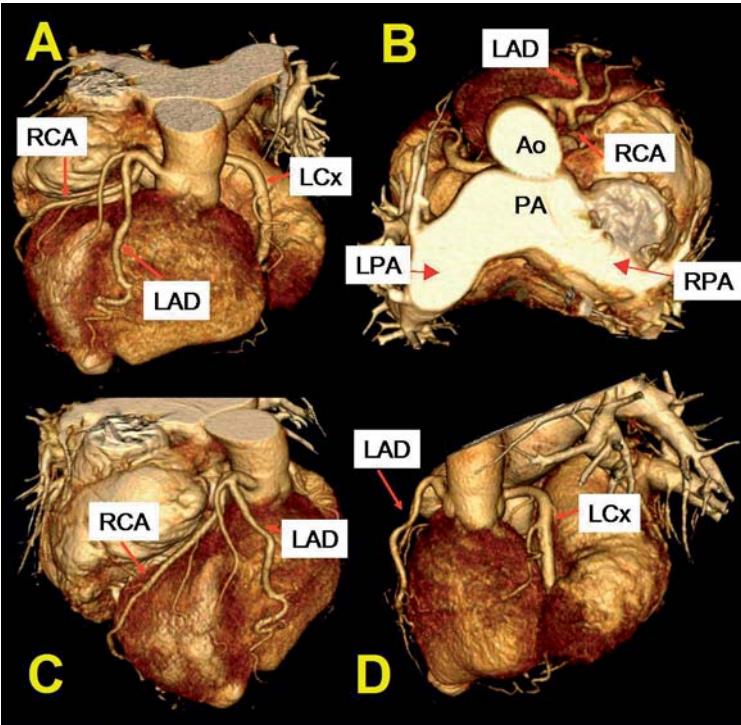


FIGURE 2.37

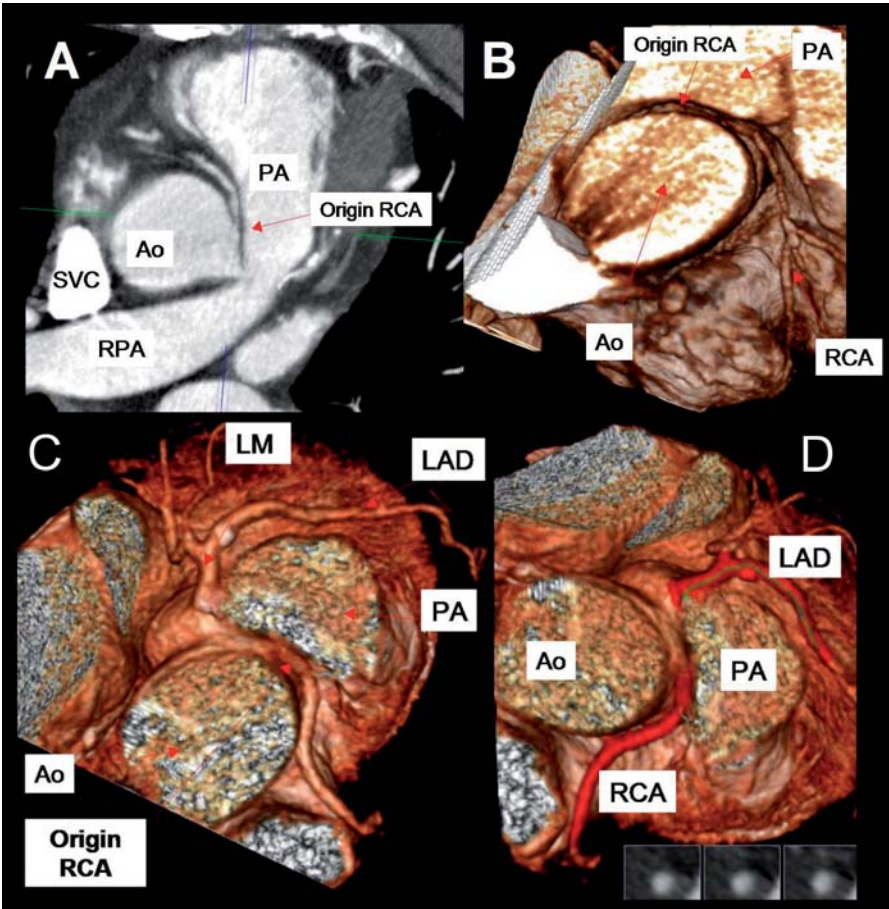


FIGURE 2.38

- F. 2.37. Patient with transposition of the great arteries and an associated anomaly of the origin and course of the coronary arteries. A (anterior view): The left anterior descending (LAD) is seen at the anterior interventricular groove; B (cranial view) and C (right anterior oblique view): The LAD and the right coronary artery (RCA) arise from a common trunk; D (left anterior oblique view): The circumflex artery (LCx) emerges independently from the posterior sinus of Valsalva; Ao: aorta; LPA: left pulmonary artery; PA: main pulmonary artery; RPA: right pulmonary artery.
- F. 2.38. Anomalous origin of the right coronary artery (RCA), emerging from the left coronary sinus of Valsalva and following an interarterial course between the aorta (Ao) and the pulmonary artery (PA) from two different patients (A,B and C,D). A: Axial oblique MPR; B-D: 3D volume rendering reconstructions (cranial view); LAD: left anterior descending; LM: left main trunk; RPA: right pulmonary artery; SVC: superior vena cava.

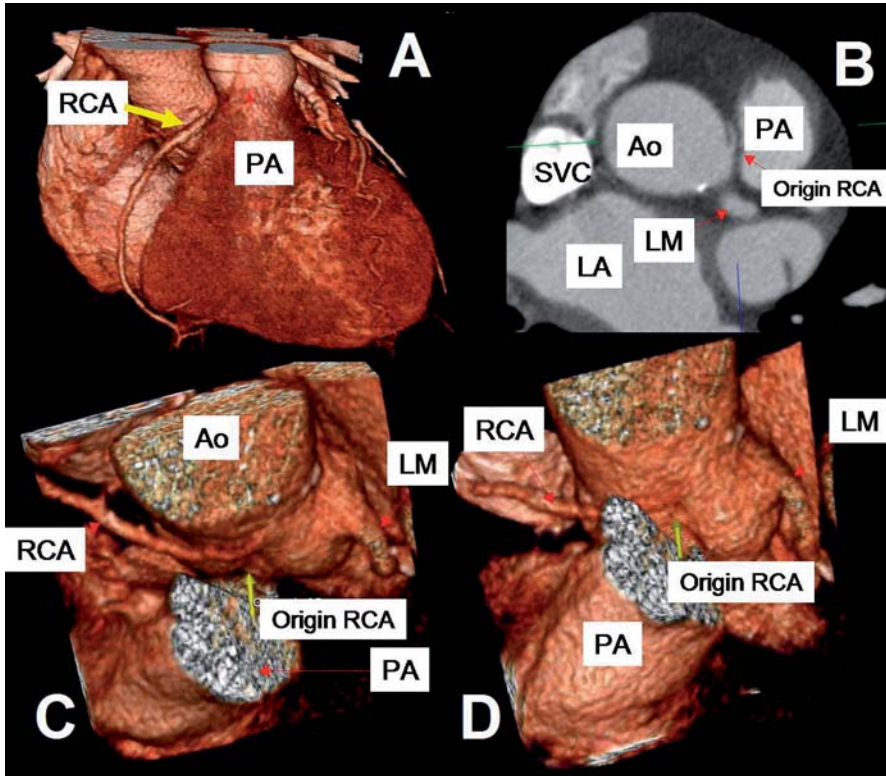


FIGURE 2.39 Anomalous origin of the right coronary artery (RCA) emerging from the left sinus of Valsalva, viewed from 3D volume rendering images (A, C, D) and oblique MPR (B), all from the same patient. Ao: aorta; LA: left atrium; LM: left main; PA: pulmonary artery; SVC: superior vena cava.

2.5.2 Anomalies not leading to myocardial ischemia

Some of the congenital coronary anomalies are not related to myocardial ischemia and are detected incidentally. However, their knowledge is important for two reasons, particularly when non-coronary cardiac surgery is planned to prevent inadvertent damage of the aberrant coronary vessel, and to appropriately guide the coronary artery cannulation^{25,37,41–43}.

These coronary anomalies have been detected in 0.5–1% of invasive coronary angiographies, although it is not always feasible to describe at angiography the exact course of the abnormal

artery in relation to the great vessels⁴⁴. MDCT has shown better diagnostic accuracy in this aspect, provided its ability to present 3D reconstructed images^{45–47}.

The most frequently found types of coronary anomalies are the following:

Origin of the LCx from the right coronary sinus or RCA (Figures 2.42–2.44): It is the most frequent anomaly in this group (0.67% of diagnostic invasive coronary angiograms)^{32,48} and it seems to be more frequent in cases of congenital stenosis of the aortic valve^{38,43}. The LCx is seen emerging posteriorly to the RCA, and then following a course inferior and

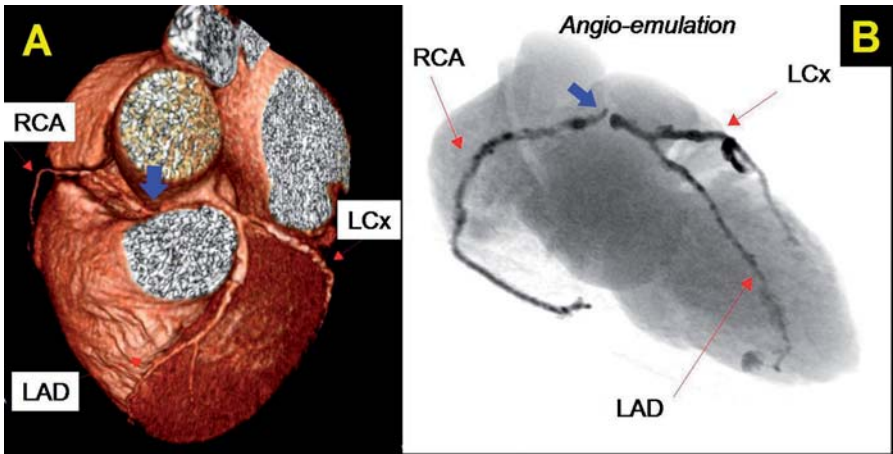


FIGURE 2.40 Anomalous origin of the right coronary artery (RCA) from the left sinus of Valsalva (blue arrows). A: 3D volume rendering; B: Postprocessed images emulating an angiography; LAD: left anterior descending. LCx: circumflex artery.

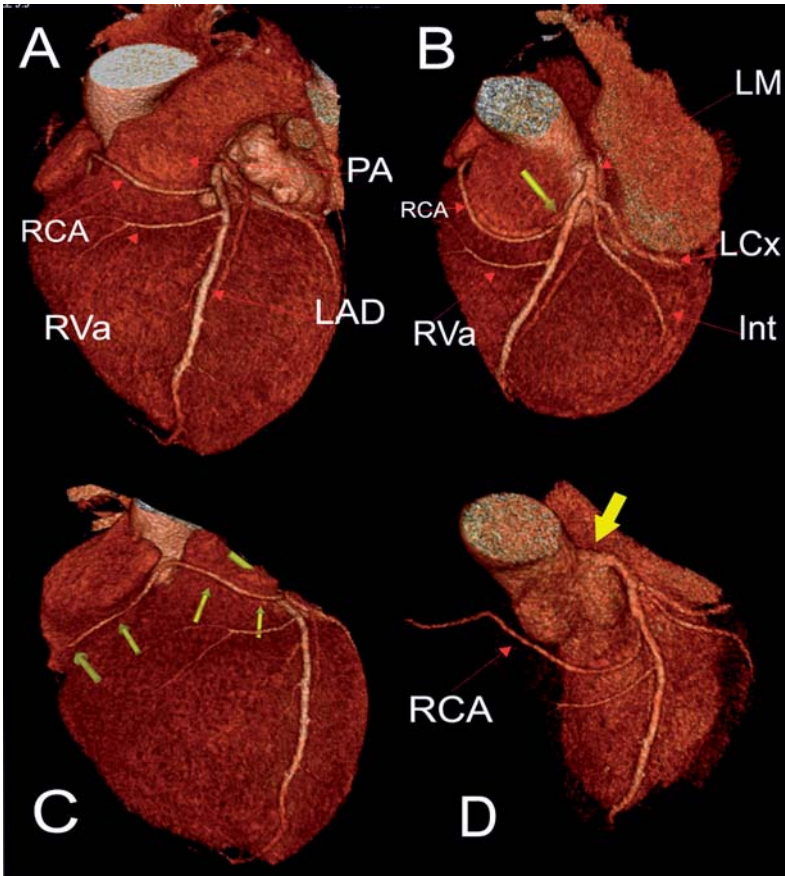


FIGURE 2.41 Anomalous origin of the right coronary artery (RCA) from the left anterior descending (LAD) coronary artery. These 3D volume rendering images correspond to the same patient, showing the origin of the RCA (A and B: yellow arrow) and the course of the vessel around the anterior aspect of the pulmonary valve ring, reaching its normal position at the right atrioventricular groove (C: yellow arrows). Distally to the abnormal origin of the RCA, the LAD gives a second abnormal branch for the right ventricle (RVa). Observe that the entire coronary arterial supply comes from a single trunk (D: yellow arrow). Int: Intermediate artery; LCx: left circumflex; LM: left main trunk.

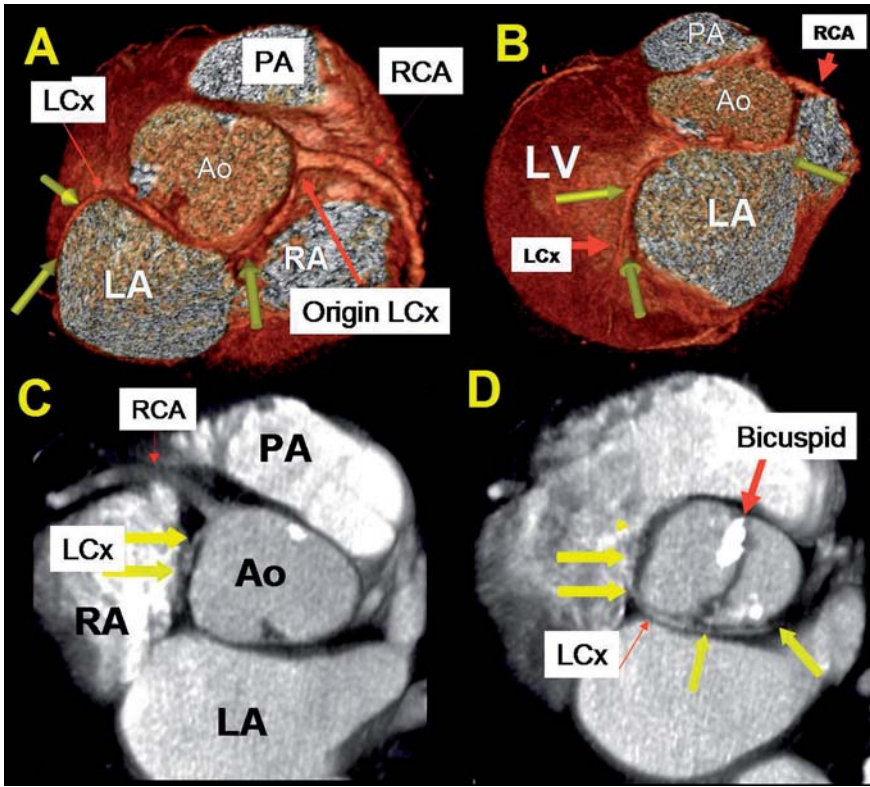


FIGURE 2.42 Patient with anomalous origin of the left circumflex artery (LCx) from the right coronary sinus of Valsalva (panels A and C), with a retroaortic course to the left atrioventricular groove (B and D). Observe the presence of a bicuspid aortic valve (panel D). Ao: aorta; LA: left atrium; LV: left ventricle; PA: pulmonary artery; RA: right atrium; RCA: right coronary artery.

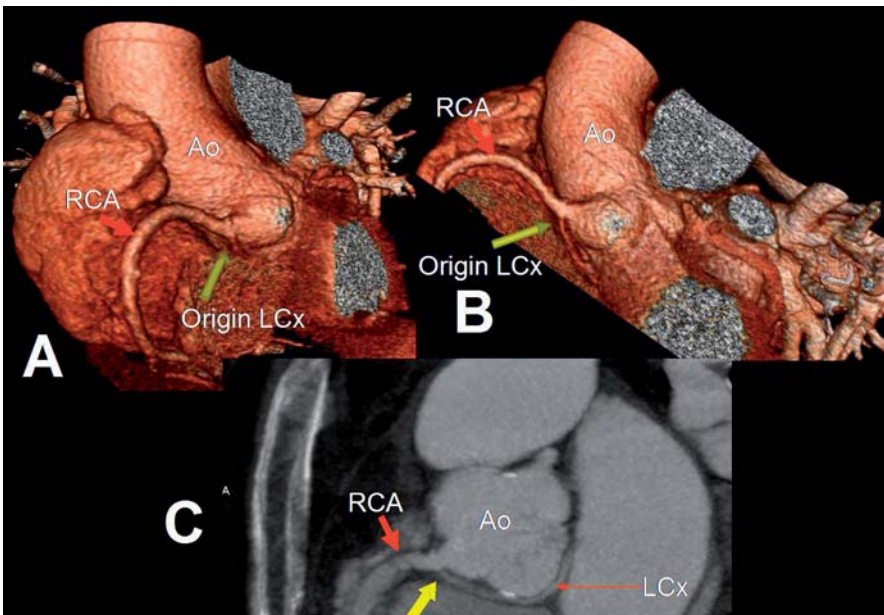


FIGURE 2.43 Patient with anomalous origin of the left circumflex artery (LCx) from the right coronary artery (RCA) (panels A and B). Observe the marked angulation of the origin of the LCx from the RCA and its retroaortic course to reach the left atrioventricular groove (panel C). Ao: Aorta.

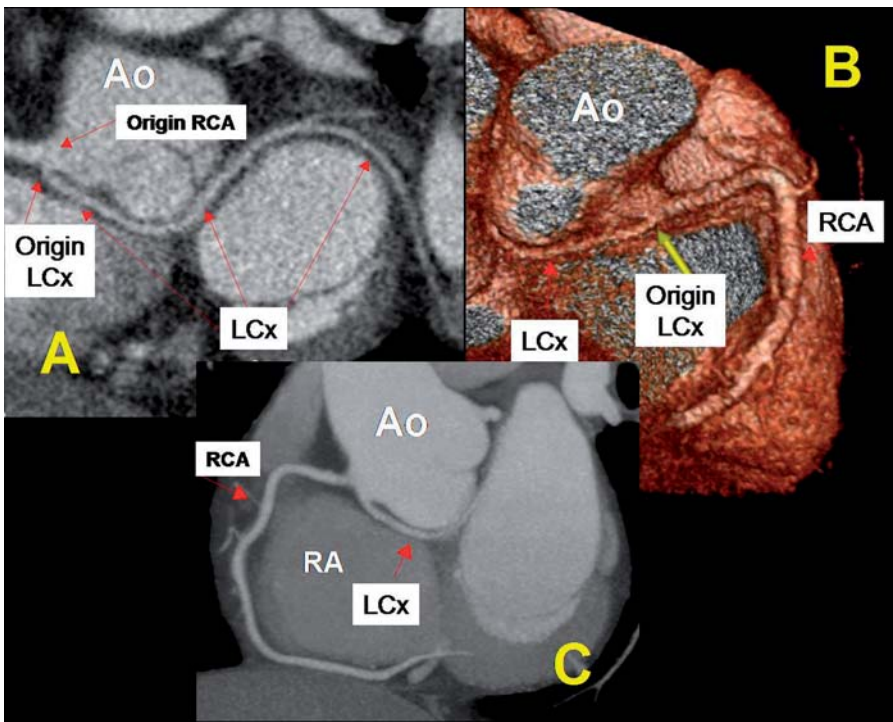


FIGURE 2.44

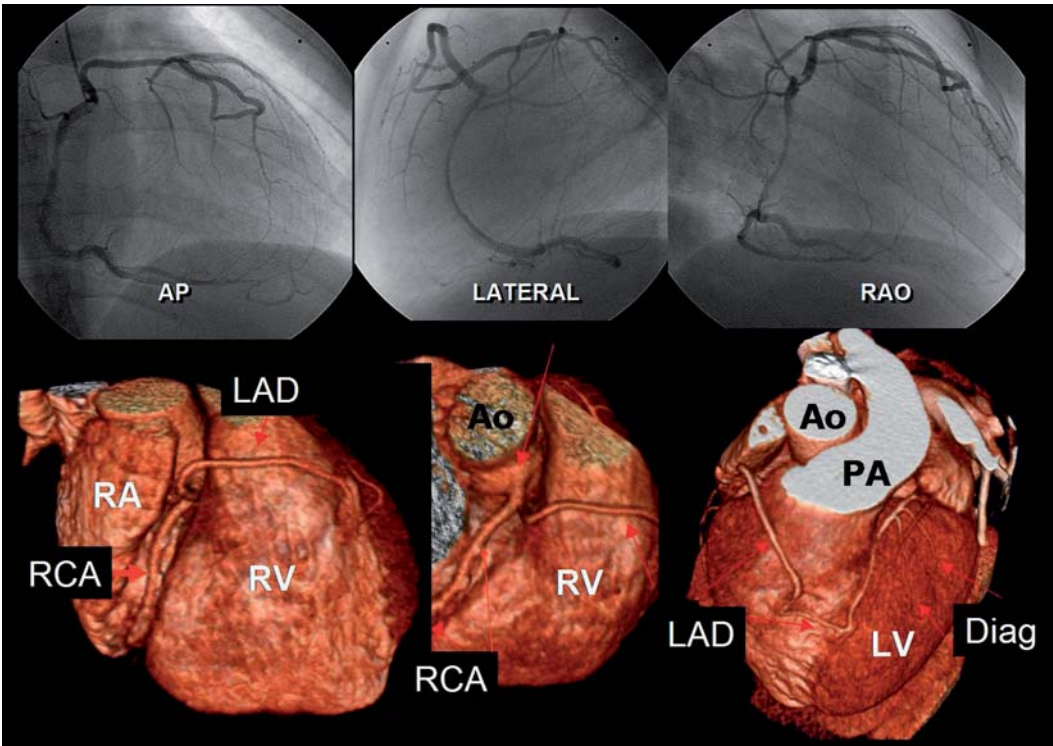


FIGURE 2.45

- F. 2.44. Examples of anomalous origin of the left circumflex artery (LCx) from the right coronary artery (RCA) (panel A: curved MPR and panel B: 3D volume rendering-, from the same patient) and directly from the right coronary sinus of Valsalva (panel C: MIP image). Ao: Aorta. RA: right atrium.
- F. 2.45. Patient with anomalous origin of the left anterior descending (LAD). Although the abnormal origin and course of the arteries is already evident on the images from the invasive angiography (top panels), the actual origin of the LAD (from the right coronary artery (RCA) or directly from the sinus of Valsalva) and their course in relation to the great vessels can only be discerned from the MDCT angiography (bottom panels). Observe the origin of the LAD from the RCA and its course over the anterior aspect of the right ventricular outflow tract, reaching the anterior interventricular groove. Ao: aorta; AP: anteroposterior; Diag: diagonal artery; LV: left ventricle; PA: pulmonary artery; RA: right atrium; RAO: right anterior oblique; RV: right ventricle.

posterior to the aortic root, reaching the left atrioventricular groove. Very rare in this case is a course of the LCx between the walls of the great vessels.

Origin of the LAD in the RCA (Figure 2.45): This anomaly is present in 4–5% of patients with tetralogy of Fallot or pulmonary atresia with ventricular septal defect²⁵. The LAD frequently shows a course anterior to the infundibular portion of the right ventricle, usually not leading to myocardial ischemia, which has potential implications in the case of surgical correction.

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Multidetector Computed Tomography

Pons-Llado, G.; Leta-Petracca, R. (Eds.)

2006, VII, 129 p., Hardcover

ISBN: 978-0-387-33044-0