Anatomic anomalies of coronary arteries: classification and evaluation using multidetector computer tomography

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Authors: A. S. Teixeira Gomes¹, M. S. R. O. Faustino², P. Campos³, I. Távora⁴, ¹PT, ²Lisboa/PT, ³Cascais/PT, ⁴Lisbon/PT
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Learning objectives

1. To illustrate and classify the multiple coronary artery anomalies studied by computed tomography (CT).
2. To access the clinical significance of each coronary artery anomaly.

Background

Coronary anomalies are present in about 1% of the general adult population [1]. Nevertheless, depending on the used definition, some studies showed higher prevalence [2]. The broad spectrum and complexity of anomalies originated multiple classification schemes. Furthermore, clinical presentation is highly variable [3], from asymptomatic patients to sudden death events. Currently, no single classification scheme is widely used. In this presentation we will opt to distinguish coronary anomalies based on its clinical significance, as this allows a more practical approach. All coronary artery anomalies should be reported since even benign anomalies may have clinical implications when associated with atherosclerotic lesions or during surgical or angiographic procedures.

Recent CT technical improvements enabled its application in the study of the coronary arteries, allowing visualization of coronary anatomy. The combination of an excellent temporal and spatial resolution and the three-dimensional visualization of vessels makes cardiac CT the standard of reference for evaluation of coronary anomalies [4]. Several studies have confirmed cardiac CT superiority when compared with the previous standard, coronary angiography [3], while also being a non-invasive modality. Magnetic resonance (MR) will prove useful to study the coronary arteries anatomy, but its role isn't as clear as cardiac CT.

Imaging findings OR Procedure details

Between May 2010 and August 2012, anatomic coronary anomalies were found in 14 patients from a non-pediatric population. CT scans were performed with a 64-slice spiral CT scanner. Contrast was administered and the acquisition start was done manually, with its timing measured from a previous bolus-tracking. Beta blockers were administered to lower the heart rate below 70 cycles per minute when necessary. Retrospective ECG gating was used to reconstruct the axial images at different percentages of R-R interval, evaluating those with the least motion artifacts for each artery. In addition to the axial
images, multiplanar reconstruction (MPR), maximum and volume intensity projection (MIP and VIP) and volume-rendered images (3D - VRI) were created.

**Hemodynamically Significant Coronary Anomalies**

1. **Ectopic Origin from Pulmonary Artery** - More frequently, the left coronary artery originates from the pulmonary artery (left posterior sinus), an entity known as Bland-White-Garland syndrome or ALCAPA (anomalous origin of the left coronary artery from the pulmonary artery). It is present in about 1/300000 newborns [5]. Symptoms usually develop during the first weeks of life due to perfusion by relatively desaturated (pulmonary) blood under low pressure. Later occurs a development of collaterals between left and right coronary systems, and subsequently a "coronary steal" phenomenon through the left coronary artery to the pulmonary artery. In patients who live to adulthood this entity may provoke myocardial infarction and left ventricular dysfunction or silent myocardial ischemia, which is a risk to sudden cardiac death [5]. Rarer variations of this syndrome can involve any other coronary branch or be associated with other congenital heart malformations.

2. **Interarterial course** - This anatomic arrangement can happen when a coronary artery (or one of its branches) arises from the contra-lateral coronary artery or aortic sinus (Fig. 1 on page 12; Fig. 3 on page 13) and it is the most common hemodynamically significant anomaly. While other courses such as prepulmonic, retroaortic or transseptal configurations usually carry no clinical significance, interarterial course is associated with sudden cardiac death, particularly in young athletes [4]. There are two possible explanations to this phenomenon [6]: compression of the artery between the aorta and the pulmonary artery (Fig. 2 on page 13 A) or occlusion due to an abnormal orifice (slitlike) (Fig. 2 on page 13 B); both aspects would be aggravated during exercise. An intramural course within the aortic wall may increase the risk of sudden death, therefore this morphologic feature ought to be reported. Left anterior descending artery or left coronary artery with interarterial courses (Fig. 3 on page 13) have been described as more prone to be clinically significant. There isn't yet a consensus when surgery should be performed since the causal link to sudden death is not fully established.
Fig. 1: Origin of the right coronary artery (RCA) from the left coronary sinus in a 70 year old man with chest pain. MIP (A) and VRI (B) images show the RCA with an interarterial course (arrow) between the aorta (Ao) and pulmonary artery (PA). This patient had a pacemaker and left anterior descending artery (LAD) stenting.

References: Centro Hospitalar Lisboa Norte - /PT

Fig. 2: Schematic representation of two possible mechanisms which explain hemodynamic consequences of interarterial course: compression (A)
between the aorta (Ao) and the pulmonary artery (PA) and occlusion (B) of the right coronary artery (RCA) orifice.

References: A. S. Teixeira Gomes

**Fig. 3:** Origin of the left coronary artery (LCA) from the right coronary sinus in a 73 year old man with previous myocardial infarction. MIP (A) and VRI (B) images demonstrate the left coronary artery originating from the right aortic sinus with an interarterial course (arrow) between the aorta (Ao) and pulmonary artery (PA). Right coronary artery (RCA).

References: Centro Hospitalar Lisboa Norte - /PT

3. Atresia - A rare condition that usually affects the left coronary artery and manifests itself in the first year of life [7], although some cases have been reported in adulthood. Collateral right-to-left circulation is often evident, albeit insufficient to properly supply blood to the left myocardium. Surgical intervention is often the treatment of choice.

4. Coronary Fistula - Fistulous communication can occur between a coronary artery and either the coronary venous system, main pulmonary artery (Fig. 4 on page 14; Fig. 5 on page 14) or the cardiac chambers, more frequently the right cardiac chambers. It is significant due to the pressure gradient between the two structures, which occurs because the normal capillary bed isn’t interposed, resulting in a steal phenomenon [6]. This may lead to the characteristic enlargement and turtuosity of the coronary artery.
Fig. 4: Fistulous communication in a 54 year old woman with heart murmur. MIP (A) and VRI (B) images show a fistulous communication (arrow) between the left anterior descending artery (LAD) and the pulmonary artery (PA). Aorta (Ao). Another fistula involving the right coronary artery in the same patient is better seen in figure 5.

References: Centro Hospitalar Lisboa Norte - /PT

Fig. 5: Fistulous communication in a 54 year old woman with heart murmur. MIP (A) and VRI (B) images show a fistulous communication (arrow) between the right coronary artery (RCA) and the pulmonary artery (PA).
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5. Myocardial bridge - Normal coronary arteries have an epicardial course, surrounded by a layer of fat. Normal arterial branches that course into the myocardium form a perpendicular angle with the parent artery. Myocardial bridge is described when the parent arteries themselves dive into the myocardium (Fig. 6 on page 15), reemerging in a subepicardial location more distally. The affected segment is called the tunneled segment. Reported prevalence is highly variable ranging from 15% to 85% in autopsy studies and 0.5% to 12% in angiographic studies [8]. Nonetheless the depiction rate of myocardial bridging with CT is greater than that of conventional angiography. Hemodynamic significance is difficult to establish and sometimes controversial: while generally benign, it has been associated with infarction and sudden cardiac death [9]. Systolic compression of the tunneled segment is the described mechanism, with some reports [8] finding correlation with depth, but not length, of the myocardial bridge. Also, atherosclerotic plaque formation is frequently accentuated in the proximal segment of the myocardial bridge. Therapeutic intervention decision should have a multidisciplinary approach in a case to case basis.
**Fig. 6**: Miocardial bridge in a 25 year old man with atypical chest pain. MIP image show a small miocardial bridge (arrow) in the left anterior descending artery (LAD).

**References:** Centro Hospitalar Lisboa Norte - /PT

**Benign Coronary Anomalies**

1. Absence of the Left Main Coronary Artery - In this condition both left anterior descendent and circumflex arteries arise separately from the left aortic
sinus. It is a relatively frequent anomaly without hemodynamic repercussion [9].

2. Prepulmonary, retroartoric and transseptal courses - As with interarterial courses it can occur when a coronary artery (or one of its branches) originates from the contra-lateral coronary artery or aortic sinus.
   - Prepulmonary course is found more frequently when the left coronary artery arises from the right aortic sinus and is a benign variant. Prepulmonary course is common in tetralogy of Fallot [6], which can complicate surgical intervention.
   - Commonly, it is the circumflex artery that has a retroartic course (Fig. 7 on page 16), rising from the right sinus, with no hemodynamically significance. However, it may complicate aortic valve surgery because a retroartic artery partially encircles the valve.

![Fig. 7](image)

**Fig. 7**: Anomalous circumflex artery in a 58 year old male with chest pain and positive cardiac stress test, found in a previous angiographic study. MIP (A) and VRI (B) images demonstrate the circumflex artery with a retroaortic course (arrow) arising from the right coronary artery (RCA) ostium. Extensive calcified plaques in both left anterior descending artery (LAD) and right coronary artery are present. Aorta (Ao).

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- Transseptal course (Fig. 8 on page 17) is distinctive from interarterial course because, in the former case, the artery runs through the septal myocardium (Fig. 9 on page 17 B). Other feature such as coursing downward (Fig. 8 on page 17 B) (hammock sign) or the absence of a slitlike orifice, help distinguish from the interarterial course. This distinction (Fig. 9 on page 17) is important since the transseptal course is considered benign [3].
**Fig. 8:** Left coronary artery (LCA) with anomalous origin from the right coronary sinus in a 48 year old man with previous myocardial infarction. MIP images show the LCA with a transseptal course (arrow) and a visible proximal downward course (arrow in figure B), the "hammock sign". Right coronary artery (RCA).

**References:** Centro Hospitalar Lisboa Norte - /PT

**Fig. 9:** Sagital oblique CT images show the difference between an interarterial course in figure A (arrow) and a transseptal course in
Coronary Duplication - Most commonly, the left anterior descendent artery is duplicated [6], with few reports of the right coronary artery also having been described. In spite this anomaly not being clinically significant, as with other benign anomalies, surgical intervention can be complicated [10].

High Origin - This anomaly is defined as origin of a coronary artery more than 1 cm above the sinotubular junction. Percutaneous procedures and some surgeries (such as aortic valve replacement) may prove more difficult due to high origin [7]. Right coronary artery high origin (which is more frequent than on the left) is associated with bicuspid aortic valve [3]; it is therefore important to screen for a high origin in the population before undergoing valve replacement surgery.

Systemic termination - It is a rare anomaly and can involve bronquial, pericardic or internal mammary/thoracic arteries, among others. Contrary to coronary fistulas there isn't a pressure gradient between the two vessels. Therefore, in this case, the coronary artery isn't enlarged and tortuous. Many systemic terminations might be missed during CT owing to small calibers; this has led to some authors suggesting a much higher frequency [10]. In the presence of atherosclerosis with a coronary stenosis, the subsequent pressure gradient change can accentuate the anomalous course.

Shepherd's Crook right coronary artery - in this variant the right coronary artery takes a tortuous upward course right after its origin (in the correct right aortic sinus). This is a relatively frequent (prevalence up to 5% [3]) variant, with no hemodynamic consequence. However, catheterization of the right coronary artery can be more difficult due to "kinking" of the right coronary artery (Fig. 10 on page 18).
**Fig. 10**: Shepherd's Crook in a 60 year old man with inconclusive cardiac stress test. MIP image shows an upward and tortuous proximal right coronary artery (RCA) course (arrow). Coronary plaques in the proximal RCA course are not suspected to be related with this variant.

**References**: Centro Hospitalar Lisboa Norte - /PT

**Images for this section:**
**Fig. 1:** Origin of the right coronary artery (RCA) from the left coronary sinus in a 70 year old man with chest pain. MIP (A) and VRI (B) images show the RCA with an interarterial course (arrow) between the aorta (Ao) and pulmonary artery (PA). This patient had a pacemaker and left anterior descending artery (LAD) stenting.

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Fig. 9: Sagital oblique CT images show the difference between an interarterial course in figure A (arrow) and a transseptal course in figure B (arrow). In both images the artery is the left coronary artery with anomalous origin from the right aortic sinus.
Fig. 10: Shepherd's Crook in a 60 year old man with inconclusive cardiac stress test. MIP image shows an upward and tortuous proximal right coronary artery (RCA) course (arrow). Coronary plaques in the proximal RCA course are not suspected to be related with this variant.
Conclusion

CT coronariography allows clear visualization of normal coronary anatomy and its most frequent anomalies. Radiologists should be familiar with the spectrum of coronary artery anomalies and the clinical significance of each anomaly to correctly interpret CT coronariography exams and even non-cardiac thoracic CT exams (Fig. 11 on page 20), where some anomalies could otherwise be missed.

Images for this section:

Fig. 11: Left coronary artery with anomalous origin from the right coronary sinus and transseptal course. Although better seen in axial cardiac CT image (B), this anomaly is clearly visible in an axial image of a non-cardiac contrasted thoracic CT (A).
References


Personal Information