

Type IV Dual Left Anterior Descending Coronary Artery Anomaly with Prepulmonic Course: Differential Diagnostic Features with Coronary Angiography: Case Report

Prepulmonik Seyir Gösteren Tip IV Çift Sol Ön İnen Koroner Arter Anomalisi: Koroner Anjiyografik Ayırıcı Tanısal Özellikler

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ABSTRACT Congenital coronary artery anomalies (CAA) are uncommon and the vast majority is diagnosed incidentally during coronary angiogram or necropsy. Their prevalence is less than 1.3% based on published series. Diagnostic difficulties and their highly variable incidence have made CAAs of great interest to cardiologists. Dual left anterior descending artery (LAD) distribution with either of the vessels originating from the left main coronary artery and the right sinus of Valsalva without associated congenital cardiac defects is an extremely rare CAA and the determination of its course (interarterial, intraseptal, prepulmonic or retroaortic) is very important regarding its clinical significance. In this report, a type IV LAD case with prepulmonic course was presented regarding its typical and differential diagnostic features with angiography.

Key Words: Coronary vessel anomalies; coronary vessels

ÖZET Konjenital koroner arter anomalileri (KAA) sık değildir ve çoğunluğu tesadüfi olarak koroner anjiyografi ya da nekropsi çalışmalarında saptanırlar. Yayımlanmış serilerde prevalansı %1,3 den azdır. Tanısal güçlükleri ve oldukça değişken insidansları nedeni ile kardiyooloji pratiğinde ilgi çekici olmuşlardır. Konjenital kardiyak defektler olmaksızın, sol ana koroner arter ve sağ valsalva sinüsünden köken alan damarların her ikisinin katıldığı LAD ayrılması (dallanması) çok nadir bir KAA olup, seyirinin (interarteriyel, intraseptal, prepulmonik ya da retroaortik) saptanmasının klinik önemi fazladır. Bu raporda, prepulmonik seyirli bir tip IV LAD olgusu anjiyografik tanısal ve ayırıcı tanısal özellikleri vurgulanarak sunulmuştur.

Anahtar Kelimeler: Koroner damar anomalileri; koroner damarlar

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Congenital coronary artery anomalies (CAA) are uncommon and the vast majority is diagnosed incidentally during coronary angiogram or necropsy. Their prevalence is less than 1.3% based on published series.^{1,2} Diagnostic difficulties, a lack of knowledge about the mechanisms through which they produce myocardial ischemia, and their highly variable incidence have made CAAs of great interest to cardiologists. However, given their low incidence, most interventional cardiologists have a limited experience in the study of CAA.³

The most common CAA is separate origination of the left anterior descending (LAD) and left circumflex (LCX) from the left sinus of Valsalva. The second most common anomaly is origination of the LCX artery from the right coronary artery (RCA) or right aortic sinus of Valsalva (RSV).^{1,2} Dual LAD distribution with either of the vessels originating from the left main coronary artery (LMCA) and the RSV without associated congenital cardiac

defects is an extremely rare CAA^{1,2} and the determination of its course (interarterial, intraseptal, prepulmonic or retroaortic) is very important regarding its clinical significance.³⁻⁵

CASE REPORT

A 63-year-old man referred for diagnostic conventional coronary angiography because of high coronary risk factors and inconclusive treadmill ECG stress test, which performed for preoperative evaluation prior to orthopedic surgery. He had only mild effort dyspnea and had a history of systemic hypertension, type 2 diabetes, obstructive sleep apnea, smoking and a family history of atherosclerotic heart disease as coronary risk factors. Physical examination was unremarkable. Standard 12-lead surface electrocardiogram and chest X-ray were normal. Cardiac catheterization was performed via the right femoral artery by means of the Judkins technique. The left ventriculogram showed a normal chamber size and wall motion. Selective injection of the contrast media into the LMCA revealed a normal left main and LCx. A short LAD originated from the LMCA and, running normally, terminated after second diagonal branches, at the

proximal segment of the anterior interventricular sulcus (AIVS) (Figure 1A). The area that is normally perfused by the distal portion of the LAD was found to be avascular and free of collateral circulation (Figure 1A). We considered at first that the RCA might be long, wrapping the cardiac apex and supplying various portions of the apical, distal AIVS segments. Selective right coronary angiography with a right coronary Judkins catheter technique revealed a normal RCA and free of distal AIVS collateral area in right anterior oblique position (RAO) (Figure 1B). However, an another vessel arising just above the origin of the RCA was noticed incidentally in left anterior oblique position (LAO) (Figure 2). This anomalous vessel took an anterior course across the right ventricular outflow tract, giving rise to prepulmonic conus artery, and then turned sharply downward to descend over the mid and distal AIVS, giving rise to several small septal branches (Figure 3A and B). These coronary angiographic findings were considered to be consistent with dual LAD distribution of the more common type IV variant Spindola-Franco and colleagues classification in which the short LAD originated from the LMCA, and the long

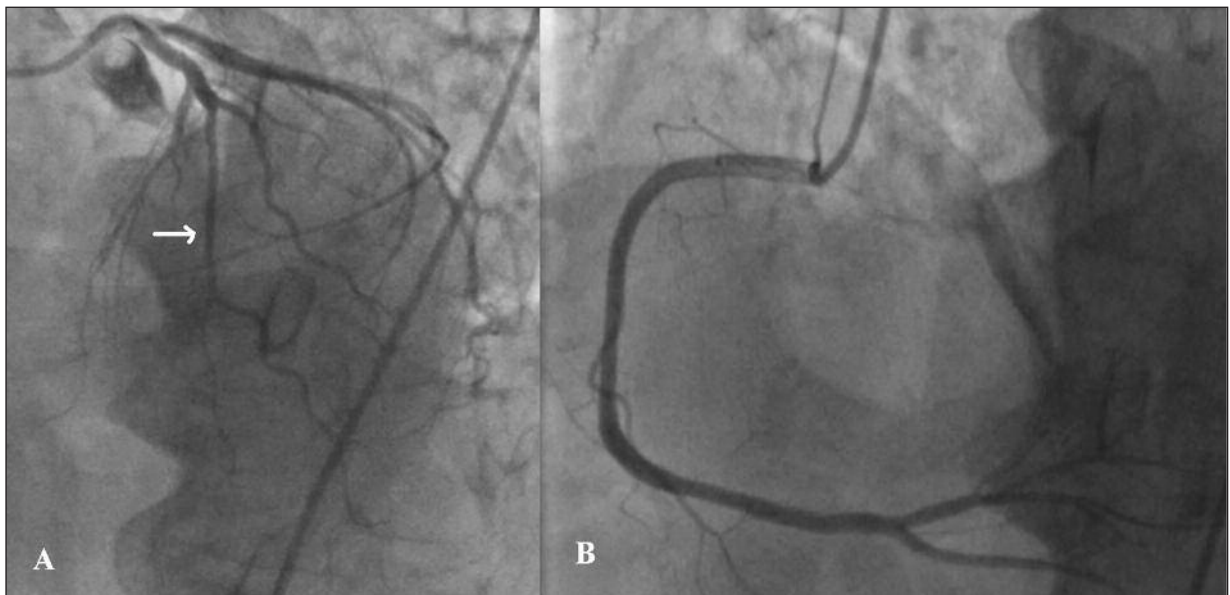


FIGURE 1: Illustrations of coronary angiographic images of the LAD, LCX (A) and RCA (B) in LAO with cranial angulation. A short LAD (arrow) originated from the left main coronary artery and, running normally, terminated after second diagonal branches, at the proximal segment of the AIVS and an unperfused myocardium of distal portion of the LAD are seen (A). The normal RCA course without distal AIVS collateral network also is seen (B). LAD, left anterior descending coronary artery; LCX, circumflex coronary arteries; RCA, right coronary artery; LAO, left anterior oblique projection; AIVS, anterior interventricular sulcus

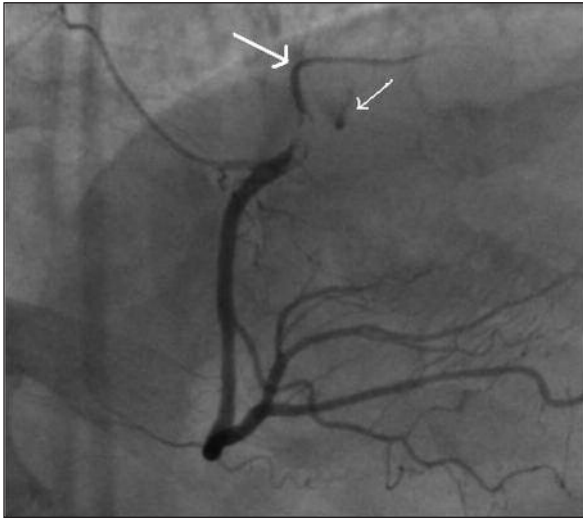


FIGURE 2: Illustrations of coronary angiographic image of the RCA in RAO. An another vessel arising just above the origin of the RCA (thick arrow) and orifice of left main coronary artery (thin arrow) are also seen in addition to normal RCA course. RCA, right coronary artery; RAO, right anterior oblique projection.

LAD originated from a separate ostium close to the orifice of the RCA with prepulmonic course.

DISCUSSION

The LAD normally originates from the LMCA, courses in the AIVS toward the cardiac apex, and

gives off diagonal branches to the anterior wall of the left ventricle and septal perforators to the interventricular septum. In rare instances, dual LAD (also called dual anterior interventricular artery) is found. This anomaly is characterized by a short LAD that terminates high in the AIVS and a long LAD that has a proximal course outside the AIVS and returns to the sulcus in its distal course. Depending on the origin of major septal, conal and diagonal branches and course of long LAD, it has been classified into 4 different subtypes by Spindola-Franco et al.⁵ Recently, the new variants of dual LAD as a type V also has been recommended.^{6,7} While the short LAD terminates in the proximal aspect of the AIVS in all types, the long LAD has a variable course outside the AIVS and returns to the inside distally, and determines the subtypes.⁵

In the most common form (type I), the short and long LADs originate from the normal proximal LAD (known as LAD proper). In the type II variety, which is very similar to type I, the short LAD is the same as in type I; however, the long LAD courses over the anterior surface of the right rather than the left ventricle, before reentering the AIVS.

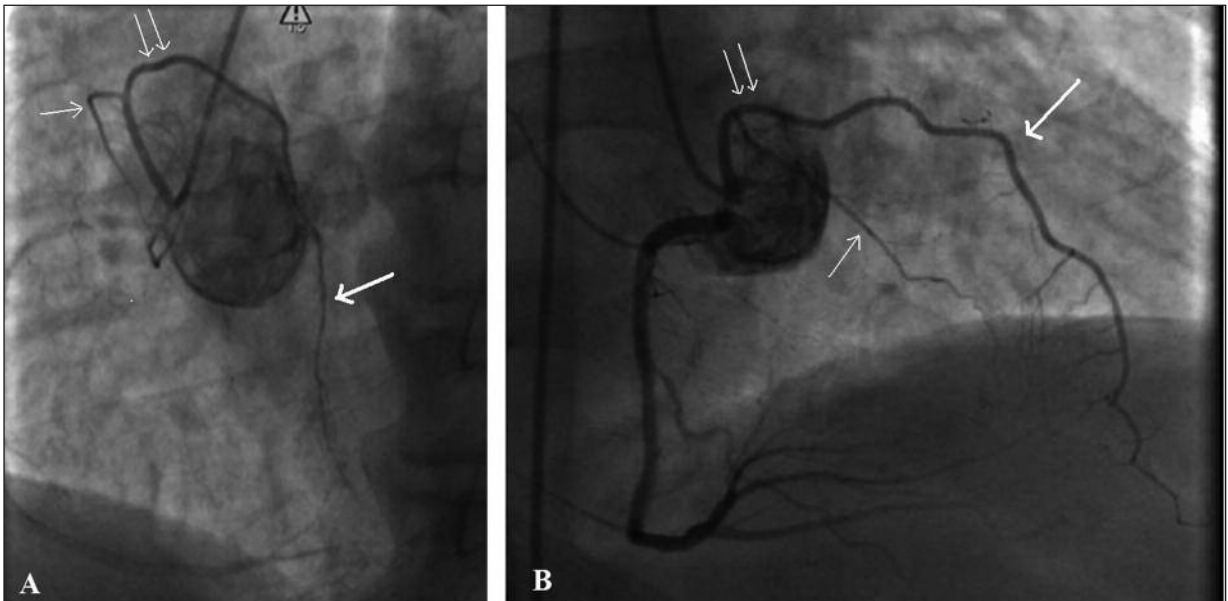


FIGURE 3: Illustrations of coronary angiographic images of the LAD in LAO with cranial angulation (A) and RAO (B). This anomalous vessel is seen to take an anterior course across the right ventricular outflow tract (thick arrow), giving rise to prepulmonic conus artery (thin arrow), and then turned sharply downward to descend over the mid and distal AIVS (anterocranial curvature, double arrow), giving rise to several small septal branches. LAD, left anterior descending coronary artery; LAO, left anterior oblique projection; RAO, right anterior oblique projection; AIVS, anterior interventricular sulcus.

In the type III dual LAD, the long artery has, at least partly, an intramyocardial (bridging) course. Unlike types I and II, the septal perforators arise from the long LAD and the diagonals arise from the short LAD. Finally, in the type IV variety, the short LAD arises from the LMCA and the long artery anomalously arises from the RSV or RCA.⁵⁻¹⁰

In general, this anomaly has no clinical significance unless atherosclerotic coronary artery disease superimposes. However, the long LAD's course is important. When the LAD originates from the proximal RCA or RSV, this anomalous LAD takes 1 of 4 aberrant pathways to reach its proper vascular territory (Table 1).^{5,10,11} These pathways are designated as type **A** (Anterior to the right ventricular outflow tract- “prepulmonic, conal or anterior free wall course with antero-cranial convex curvature”), type **B** (Between the aorta and pulmonary trunk- “interarterial course with postero-cranial convex curvature”), type **C** (Cristal, coursing through the crista supraventricularis portion of the intramuscular course through the septum, along the floor of the right ventricular outflow tract – “intraseptal course with antero-caudal concave curvature”), and type **D** (Dorsal or posterior to the aorta- retroaortic course with postero-caudal concave curvature and the “aortic root” or Page’s sign) (Table 1).^{3,5,11-13} In the report by Tuncer et al., this course of accessory LAD anterior to the right ventricular outflow tract accounted for 67% of their patients with dual LAD.¹⁰

Our case had a “prepulmonic LAD” anomalously and was consistent with type IV dual LAD CAA of this classification, which is the less frequent type. The long LAD did not originate from RCA, but from the RSV with an independent ostium and had an antero-cranial convex curvature.

The course of the long LAD can invasively be identified by angiography after insertion of a catheter into the pulmonary artery and another into the aorta,⁹ noninvasively by MSCT¹⁴⁻¹⁷ or by the “dot and eye method” in coronary angiography.^{11,12} In 1985, Ishikawa et al.¹¹ followed by Serota et al in 1990¹² proposed a series of coronary arteriographic criteria intended to define the relationship, along the initial course, between the anomalous vessel and the aorta and the pulmonary artery.^{3,11,12} These authors attempted to identify the different courses on the basis of the orientation of the “loop” formed by the anomalous coronary artery with the rest of the coronary tree¹¹ or by applying the concept of the “dot” (visualization of the contrast column of the CAA which, because of the turn it makes, adopts the morphology of a dot.¹² In a 30° RAO view, before turning to the apex at the midseptum, the LAD will pass left and upward (forming the upper half of the “eye”- antero-cranial convex loop-curvature) in an anterior free wall, prepulmonic or conal course, while it will pass left and downward (forming the lower half of the “eye”- antero-caudal concave loop-curvature) in an intraseptal course. In case of an interarterial course the vessel will describe a cranial posterior

TABLE 1: The subtypes depending on the aberrant pathways to reach its proper vascular of territory of the anomalous LAD.

Type	Definition	Course	Curvature of long LAD	Clues
A	Anterior to the right ventricular outflow tract	Prepulmonic (known as also precardiac)	Antero-cranial convex	If the first branch arising from the long LAD is the conal or infundibular branches
B	Between the aorta and pulmonary trunk	Interarterial (known as also preaortic)	Postero-cranial convex	
C	Cristal, coursing through the crista supraventricularis portion of the intramuscular course through the upper portion of the intraventricular septum	Intraseptal (known as also subpulmonic or septal)	Antero-caudal concave	If the first branch arising from the long LAD is the septal branch
D	D-orsal or posterior to the aorta	Retroaortic (known as also retrocardiac)	Postero-caudal concave	Aortic root sign or Page’s sign

loop before turning anteriorly towards the AIVS in a RAO or LAO views (postero-cranial convex curvature). The intraseptal course is also favored by the presence of phasic systolic compression of this segment similar to a myocardial bridge and proximal branching into septals. Failure to recognize such a branching of an aberrant LAD may lead to misinterpretation of an intraseptal course for a prepulmonic.¹⁸ To avoid this misinterpretation, it is important to define the a branching of an aberrant long LAD correctly as a septal artery (in allways intraseptal course) or a conus artery (prepulmonic-conal course) despite both may have anterocranial curvature-loop. Recognition of this variant and differentiation among the possible courses of the ectopic branch is essential for patient management.^{13,18,19} Because, the LAD must be coursing inside the septum, if a septal branch arises from it (it cannot come from a prepulmonic LAD, located on the free wall of the right ventricle). Any anormal vessel that crosses from an ectopic origin on the right and goes to the left, while providing as its first branch a septal branch, has always an intraseptal course, never an epicardial prepulmonic one.^{13,18} If the first branch arising from the long LAD is the

conal or infundibular branch, which courses anteriorly to supply the muscular right ventricular outflow tract or infundibulum, this has prepulmonic course. Unfortunately, coronary angiography provides planar and not tridimensional imaging and it fails to identify at all times the anatomic context of the coronary arteries: only computerized axial tomography or magnetic resonance angiography is able to illustrate the details of those relationships.^{14,19}

It is important for both angiographers and surgeons be aware of the anatomic variants of this rare coronary anomaly, because accurate identification and delineation of the coronary arteries in the presence of coronary artery disease is integral to proper surgical or percutaneous revascularization of the myocardium, particularly in the presence of plan of corrective congenital heart surgery. A paucity of distribution of vessels in the apical LAD territory with a small LAD proper and a nonperfused distal area without collaterals during angiography of the left coronary system (the “unperfused myocardium” sign) should alert the angiographer to this as one of the likely possibilities.

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