

Coronary Artery Anomalies and Associated Radiologic Findings

Charbel Saade, PhD,* Rayan Bou Fakhredin, BSc,* Bassam El Achkar, MD,* Diamond Ghieh, MD,* Ahmad Mayat, MD,† Antoine Abchee, MD,‡ Marwan Refaat, MD,‡ Hussein Ismail, MD,‡ Hebah El-Rayess, MD,‡ Lina Karout, MD,§ and Fadi El Merhi, MD*

Abstract: Coronary anomalies occur in about 1% of the general population and in severe cases can lead to sudden cardiac death. Coronary computed tomography angiography and magnetic resonance imaging have been deemed appropriate for the evaluation of coronary anomalies by accurately allowing the noninvasive depiction of coronary artery anomalies of origin, course, and termination. The aim of this article is to describe and illustrate a comprehensive array for the classification of coronary artery anomalies.

Key Words: coronary anomalies, coronary angiography, anatomy, magnetic resonance imaging, cardiac vasculature

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Normal variants of coronary arteries display only minor deviations in their origin and course and have no associated morbidity or mortality. Approximately 1% of adults have anomalous coronary arteries that differ markedly from normal patterns. Twenty percent of congenital coronary anomalies are symptomatic or have the potential for serious sequelae such as exercise-induced arrhythmias, exertional syncope, cardiac arrest, or myocardial infarction.^{1–4} Congenital coronary anomalies have been associated with complex cardiac lesions such as transposition of the great arteries, tetralogy of Fallot, pulmonary atresia, hypoplastic left heart syndrome, Shone complex, and aortic valve disease.^{5,6} The aim of this article is to describe and illustrate a comprehensive array for the classification of coronary artery anomalies into those of origin and course, intrinsic arterial anatomy, and termination.

ANOMALIES OF ORIGATION AND COURSE

Anomalies of coronary ostia origination are considered primary anomalies, as they can exist in the absence of additional cardiac abnormalities and are not caused by coexisting anomalies. Anomalies of origination and course are a known etiology of sudden cardiac death and may be associated with any of the major coronary arteries (Figs. 1, 2).

From the *Diagnostic Radiology Department, American University of Beirut Medical Center, Beirut, Lebanon; †Diagnostic Radiology Department, Campbelltown Public Hospital, Australia; ‡Cardiology Department, American University of Beirut Medical Center; and §Department of Health Sciences, American University of Beirut, Beirut, Lebanon.

Correspondence to: Fadi El Merhi, MD, Diagnostic Radiology Department, American University of Beirut Medical Center, PO Box 11-0236 Riad El-Solh, Beirut, 1107 2020, Lebanon (e-mail: fe19@aub.edu.lb).

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Absent LMT (Split Origination of the LCA)

Because of a lack of universally accepted terminology for an absent left main trunk (LMT),^{7,8} 2 suggested criteria exist. These include the presence of 2 well-separated ostia for the left circumflex (LCX) artery and the left anterior descending (LAD) artery (Fig. 3A) or the absence of a proper LMT. Separate but adjacent ostia are usually seen in the left coronary sinus of Valsalva. The respective course of each artery is otherwise normal. Typically, there is no significant observable impairment in cardiac function.⁹ However, an increased incidence of this anomaly has been observed in hearts with left coronary artery (LCA) dominance or aortic valve disease.¹⁰

An absent LMT can be viewed on coronary angiography (CAG) with either anterior or posterior angulation of the catheter on left anterior oblique (LAO) projection.¹ It may be difficult to distinguish between a short main LMT and an absent LMT using CAG. Misinterpretation of this anomaly as either an absence or obstruction of the LCX if the catheter is positioned anteriorly or of the LAD if the catheter is positioned posteriorly in the left sinus of Valsalva can also occur with CAG.¹ In addition, the exact 3-dimensional (3D) course of an anomalous artery is difficult to ascertain with CAG because its relation to other structures is not known.¹¹ One study⁹ found that the use of multislice computed tomography allows visualization of both the proximal and distal parts of the anomalous LAD and LCX arteries in patients with absent LMTs.

Anomalous Location of Coronary Ostium Within Aortic Root or Near Proper Aortic Sinus of Valsalva

The coronary ostia mark the origin of the coronary arteries from the aorta. The normally present pair of ostia is believed to form early on during the embryologic development of the coronary circulation, just after truncal septation in the seventh to eighth week of development. The ostia are usually located in the center of the left and right aortic sinuses of Valsalva, near the free edge of the aortic cusp. This strategic location allows ample opportunity for the coronaries to fill during diastole. Anomalous located ostia can be high, low, or commissural within the aortic root or near the proper aortic sinus of Valsalva.

High Takeoff

A high takeoff coronary artery is present when the right or left coronary ostium arises 1 cm or more above the sinotubular junction, also known as the aortic cusp margin. Sudden cardiac death due to poor coronary filling and subsequent hypoperfusion and ischemia has been noted in nonathletes with high takeoff coronary.¹² It may be difficult to cannulate these patients during coronary catheterization.¹³

Alternatively, a high takeoff may be acquired secondary to proximal aortic ectasia, with dilation of the aortic root and cystic medial necrosis of the aorta. As elongation of the aorta occurs,

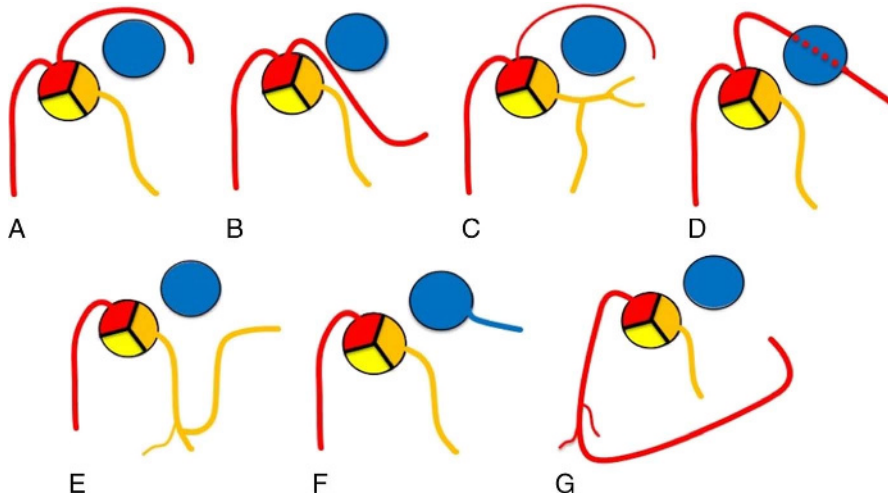


FIGURE 1. Possible anomalies of LAD coronary artery. A, Origin of the LAD from the right sinus of Valsalva, anterior course; no clinical relevance. B, Origin of the LAD from the right sinus of Valsalva, interarterial course; malignant. C, Origin of distal branches of the LAD from the right sinus of Valsalva, anterior course; no clinical relevance. D, Origin of the LAD from the right sinus of Valsalva, septal course; malignant. E, Origin of the LAD from branches of the CX; no clinical relevance. F, Bland-White-Garland syndrome of the LAD, origin from the PA; malignant. G, Origin from the LAD from the distal RCA; no clinical relevance. Figure 1 can be viewed online in color at www.jcat.org.

one or both of the coronary ostia are carried upwards, leading to acquired high takeoff.¹⁴

Ectopic coronary origin from the ascending aorta should be suspected when the coronary artery cannot be found within the sinuses of Valsalva on angiographic exploration.¹ The orifice is usually located higher than normal, within the first 2 cm of the ascending aorta. With aberrancy of the right coronary artery (RCA), the proximal segment of the vessel appears vertical in orientation when viewed from a LAO projection on CAG.¹

Commissural

The coronary ostium may also be located near the aortic valve commissure. A coronary ostium is considered *commissural* when it is within 5 mm of the commissure between 2 aortic valve

cusps. A study¹⁵ found this anomaly to be a cause of sudden death when associated with small proximal coronary arteries and the presence of a triangular septum between the upper portion of the commissure and the aorta.

Low Takeoff

Coronary arteries with a low takeoff point are found deeply hidden in the aortic sinus, immediately adjacent to a semilunar valve attachment.⁸ The anomalous coronary is more often found in the right aortic sinus than the left. It is important to identify the exact location of the takeoff before aortic valve replacement to prevent iatrogenic narrowing of the ostium or surgical occlusion with sutures, both of which have the potential to cause myocardial ischemia and infarction. Low-lying left main coronary artery

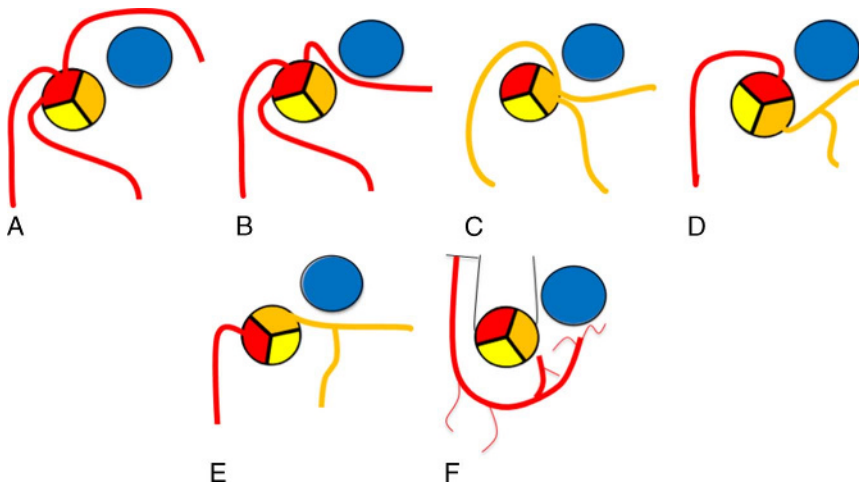


FIGURE 2. Other complex coronary anomalies. A, Origin of the RCA, CX, and LAD from the right sinus of Valsalva, anterior course of the LAD; no clinical relevance. B, Origin of the RCA, CX, and LAD from the right sinus of Valsalva, interarterial course of the LAD; malignant. C, Origin of the RCA, CX, and LAD from the left sinus of Valsalva, interarterial course of the RCA; malignant. D, Clockwise rotation of the aortic root with potential interarterial course of the RCA; depending on the extent of the malrotation, potentially malignant. E, Counter-clockwise rotation of the aortic root with a potential interarterial course of the LMA; depending on the extent of the malrotation, potentially malignant. F, Single coronary artery, ectopic origin, for example, out of the brachiocephalic trunk. Figure 2 can be viewed online in color at www.jcat.org.

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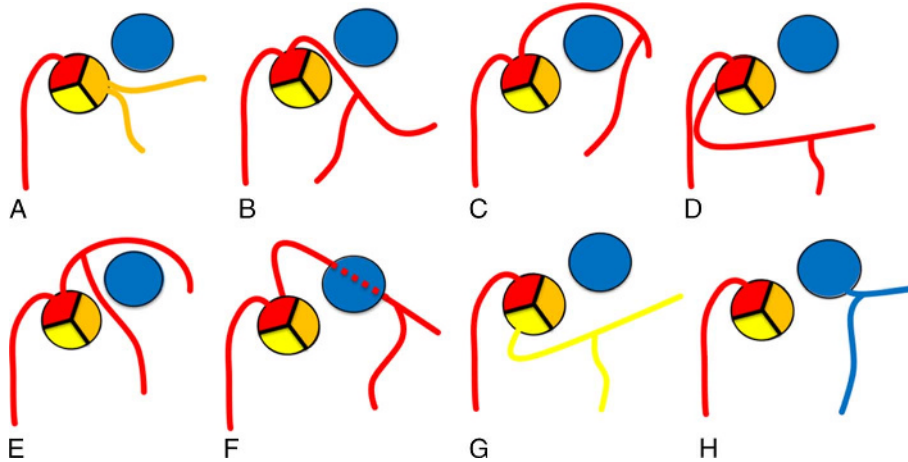


FIGURE 3. Possible anomalies of the LMCA. A, Separate ostia of the LAD and LCX from the left sinus of Valsalva; no clinical relevance. B, Origin of the LMA from the right sinus of Valsalva, interarterial course; malignant. C, Origin of the LMA from the right sinus of Valsalva, anterior course; no clinical relevance. D, Origin of the LMA from the right sinus of Valsalva, retroaortic course; no clinical relevance. E, Origin of the LMA from the right sinus of Valsalva, anterior course of the CX; malignant. F, Origin of the LMA from the right sinus of Valsalva, septal course; malignant. G, Origin of the LMA from the coronary sinus of Valsalva, no clinical relevance. H, Bland-White-Garland syndrome, origin of the LMA from the PA; malignant. Figure 3 can be viewed online in color at www.jcat.org.

(LMCA) can be seen on computed tomography 6.3 mm from the aortic annulus.

Anomalous Location of Coronary Ostium Outside Normal Coronary Aortic Sinuses

The LCA normally arises from the left posterior aortic sinus, and the RCA normally arises from anterior aortic sinus. Documented positions of anomalous ostia outside of the sinus of Valsalva have varied from the right posterior aortic sinus to the descending thoracic aorta.¹⁶

RIGHT POSTERIOR AORTIC SINUS

Ostia located in the posterior (noncoronary sinus), such as the right posterior aortic sinus, are not considered normal variants. Often the only clue to their presence is mild episodic myocardial ischemia. Both the right and left coronary arteries may aberrantly originate from the right posterior aortic sinus (Figs. 3G, 4D). Origin of the LCA from the right posterior aortic sinus (Fig. 5) has been found in association with transposition of the great arteries, whereas aberrant origin of the RCA from the noncoronary sinus (Fig. 6) has been found in association with situs inversus.

Ascending Aorta

See “Anomalous Location of Coronary Ostium Within Aortic Root or Near Proper Aortic Sinus of Valsalva” (high takeoff).

Left or Right Ventricle

Ostia may be ectopically located in either ventricle. Reverse flow from the ectopic coronary into the ventricle, known as coronary steal, occurs during both systole and diastole when the right ventricle (RV) is the point of coronary origin but only during diastole when the left ventricle (LV) is involved.¹⁷ This torrential blood flow into the ventricle may eventually lead to functional impairment.^{17,18} Collateral flow from the contralateral coronary artery is usually present in both the left and right versions of this anomaly.

Ectopic right ventricular ostia often occur in conjunction with pulmonary atresia and an intact intraventricular septum,¹⁹

in which case at least 1 coronary artery ectopically originates from the RV. Coronary blood flows retrogradely into the aorta and antegradely to supply the myocardium, often resulting in clinical ischemia. Ostial membranes, such as dysplastic aortic leaflets, may partially or totally occlude ectopic ostia originating from the LV. Often the membranes are of unclear nature.¹⁷ Direct catheter angiography usually fails to establish the locations of ventricular coronary ostia. However, CT and magnetic resonance (MR) imaging can accurately visualize these anomalies.¹⁷

Pulmonary Artery

Several variants of ostia within the pulmonary arteries have also been described and may be associated with malignant evolution. The left coronary has been observed to originate from the posterior facing sinus of the pulmonary artery (PA), as has the circumflex (CX) artery and LAD artery. Similarly, the RCA may arise from the anterior right facing sinus.

Anomalous origin of the LCA from the pulmonary artery (ALCAPA) is a rare yet serious condition almost always associated with resting ischemia, owing to reduced antegrade flow through the LCA, as well as an increased risk of fixed coronary atherosclerotic disease.¹⁶ Less frequently, ALCAPA may occur in conjunction with secondary aortic valve disease, ischemic cardiomyopathy (hibernation), volume overload, and cardiomegaly.^{16,20} Bland-White-Garland syndrome is a variant of ALCAPA where the LCA arises from the pulmonary trunk and the RCA arises from the aorta.

As early as age 1 to 2 months (infantile stage), ALCAPA may present with severe left heart failure and mitral valve insufficiency.²⁰ Those who develop sufficient collateral blood supply from the RCA may have only subtle symptoms, if any.²¹ This is known as the transition stage. The adult stage is characterized by a continuous murmur from the arteriovenous shunt into the PA.²⁰

The ALCAPA may be demonstrated most commonly originating from the left inferolateral aspect of the main PA just beyond the valvular level on MR imaging or CT.²² The reverse flow of blood into the pulmonary from the LCA can be visualized with either navigator-gated 3D whole-heart steady-state free precession sequences or phase-contrast velocity-encoded MR imaging.²²

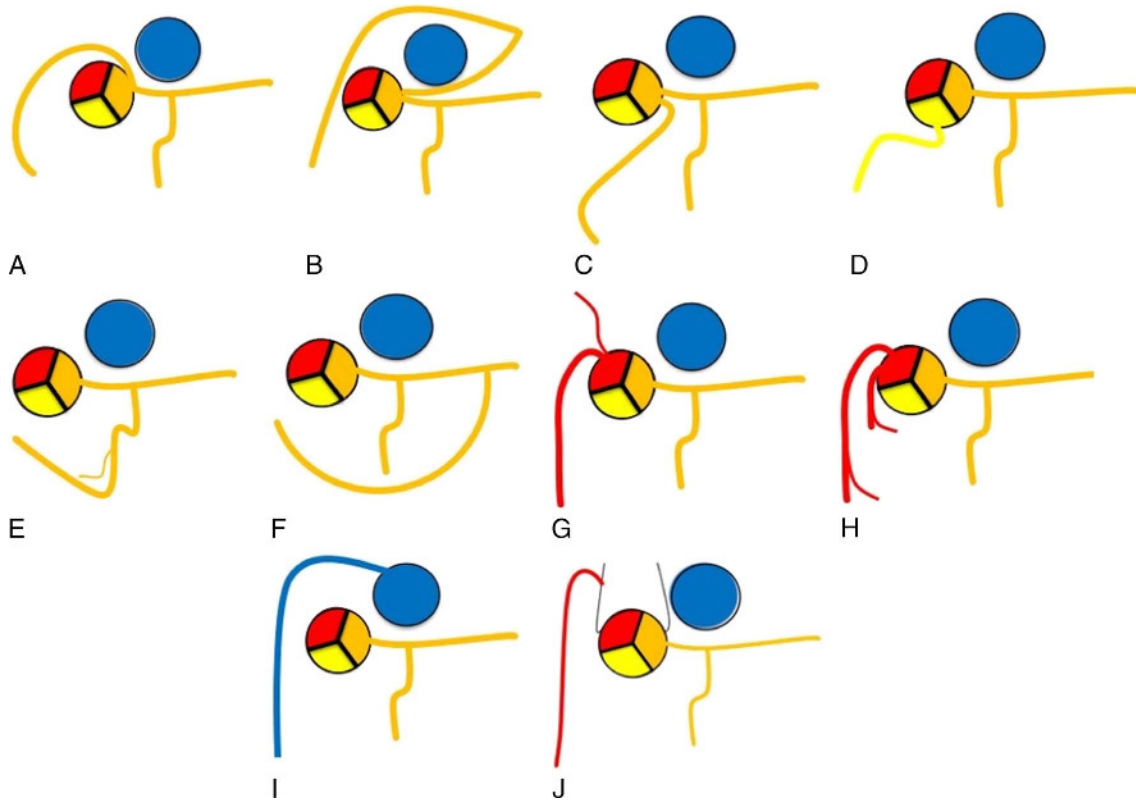


FIGURE 4. Possible anomalies of the RCA. A, Origin of the RCA from the left sinus of Valsalva, interarterial course; malignant. B, Origin of the RCA from the left sinus of Valsalva, anterior course; no clinical relevance. C, Origin of the RCA from the left sinus of Valsalva, retroaortic course, no clinical relevance. D, Origin of the RCA from the coronary sinus of Valsalva; no clinical relevance. E, Origin of the RCA from branches of the CX; no clinical relevance. F, Origin of the RCA from branches of the LAD, no clinical relevance. G, Sinus node artery with its own ostium; no clinical relevance. This anomaly is found in up to 45% of patients undergoing catheter angiography; thus, it could be considered a *normal variant*. H, Dominant conus artery (CA) with separate ostium; the CA supplies the main segment of the RCA, which is a small vessel, with an early division. The reverse situation is also possible: separate ostia of the CA and RCA, but the CA is the smaller vessel. I, Origin of the RCA from the PA, so-called reversed Bland-White-Garland syndrome; malignant. J, Ectopic origin of the RCA from the aorta, the brachiocephalic trunk, or the right common carotid artery; no clinical relevance. Figure 4 can be viewed online in color at www.jcat.org.

Imaging of adults with ALCAPA may reveal prominent collateral vessels and dilated tortuous coronaries due to shunting from the RCA to the LCA and then to the PA as well.²²

The anomalous origin of the RCA from the pulmonary artery (ARCAPA) is far less common and causes less severe signs and symptoms.²⁰ Unlike ALCAPA, ARCAPA is usually asymptomatic until adulthood, owing to the presence of adequate intercoronary collaterals that are able to maintain sufficient perfusion of the RCA territory. Roughly 50% of cases of ARCAPA are associated with other congenital heart defects such as aortic stenosis, aortopulmonary window, ventricular septal defect, and tetralogy of Fallot.²³ Traditional angiography with selective LCA angiography can be used to confirm cases of ARCAPA suspected on Doppler. Findings of angiography are filling of the RCA through collaterals followed by filling of the PA. Three-dimensional CT reconstruction imaging is useful for viewing the entire course of the RCA and for evaluating the degree of shunting.²⁴

Left anterior descending or CX arteries with PA origination are more rare than ARCAPA. Using x-ray coronary angiography, retrograde filling of the anomalous LCX via collaterals can be seen. Cardiovascular MR can be used to locate the exact origin of the anomalous LCX. An anomalous LAD originating from PA differs from adult-type ALCAPA in that no other coronaries originate from the PA and that the LCX and RCA from the aorta supply the LAD with collaterals.

Anomalous Location of Coronary Ostium at Improper Sinus (Which May Involve Joint Origination or Single Coronary Pattern)

The RCA normally originates perpendicular to the right anterior sinus of Valsalva, although several anomalous origins have been documented. The most hemodynamically significant of these is an RCA that arises from the left anterior sinus, with an anomalous route between the aorta and the PA (Fig. 4A). Such vessels are often intramural (within the aortic wall) and have acute take-off angles and thus abnormally shaped ostia (Fig. 7). The observation of sudden death in these patients is likely owing to the aortic expansion that occurs during exercise, which compresses or kinks the vessel, leading to myocardial ischemia.²⁵ Myocardial infarction, angina pectoris, syncope, ventricular tachycardia, and sudden death are rare clinical sequelae.^{1,3,25-29}

When aberrantly originating from the right aortic sinus (of Valsalva), the LCA's course turns abruptly to pass between the aorta and the right ventricular outflow tract (Figs. 3B, 8; 3C, 9). Transient occlusion occurs by a similar mechanism as described in anomalous origin of the right coronary from the left sinus; however, anomalous LCA origin from the right sinus of Valsalva is associated with higher rates of sudden death than its RCA counterpart. Using traditional angiography, the course of the anomalous coronary is best assessed with a right anterior oblique (RAO) view.³⁰

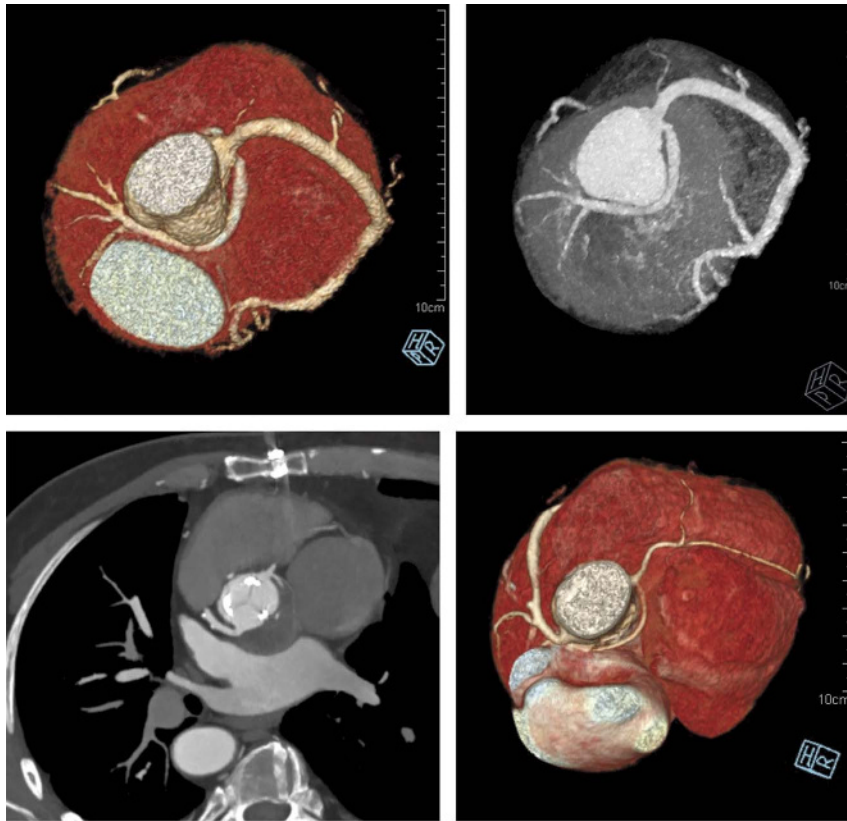


FIGURE 5. Origin of the LMA from the coronary sinus of Valsalva, no clinical relevance. Figure 5 can be viewed online in color at www.jcat.org.

Aberrant origination of the CX artery from the right sinus of Valsalva or from the most proximal segment of the RCA is the most common congenital coronary anomaly (Fig. 10A).^{20,31} The LCX artery may share the ostium of the RCA, it may arise as a proximal branch of the RCA, or it may originate from a separate ostium in the right aortic sinus.³¹ The most common route for the aberrant LCX is to course inferiorly and posteriorly from the area posterior to the RCA toward the aorta, to enter the atrioventricular (AV) groove. The vessel may have an acute take off in a retroaortic direction, between the aorta and the atrial wall. The LCX then merges in the AV sulcus. Thus, there appears to be no risk of extrinsic compression and no effect on myocardial perfusion.³¹

Single Coronary Artery

A single coronary artery is a rare anomaly where the aortic trunk has only 1 coronary ostium, which gives rise to a single coronary artery responsible for oxygenating the entire heart. The lone artery may arise from the posterior, left, or right sinus of Valsalva, with a wide range of branching pathways including intraseptal, interarterial, retrocardiac, retroaortic, and anterior courses.

Single coronary artery is less prevalent as an isolated finding than it is in association with other congenital anomalies, such as persistent truncus arteriosus³²⁻³⁴ or tetralogy of Fallot or pulmonary atresia. In some severe cardiac malformations, the single coronary artery may originate from the pulmonary trunk. This is quite

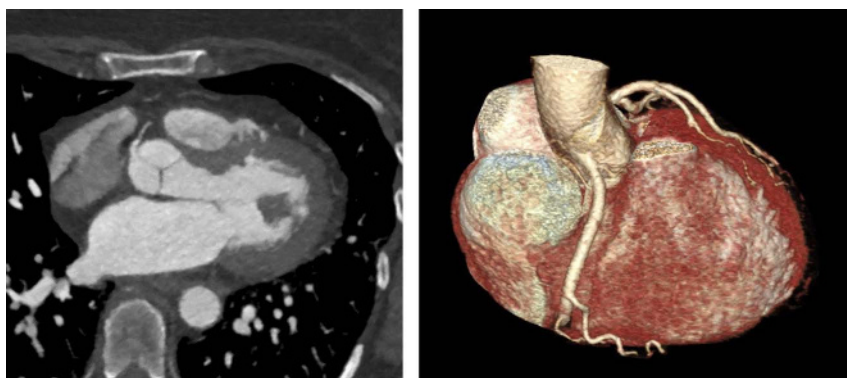


FIGURE 6. Origin of RCA from the noncoronary sinus; no clinical relevance. Figure 6 can be viewed online in color at www.jcat.org.



FIGURE 7. Origin of the RCA from the left sinus of Valsalva, interarterial course, malignant. Figure 7 can be viewed online in color at www.jcat.org.

rare, however.^{32,35–37} Patients with single coronary artery are predisposed to atherosclerotic changes owing to decreased possibility of collateralization.³⁸

The presence of only 1 coronary artery is a congenital anomaly with variable symptoms and prognoses, depending on its route. Many variations of this anomaly have been reported and are organized into the Lipton classification scheme. This scheme divides the possible courses of single coronary arteries into 3 groups. Group I consists of cases with vessels that follow the normal course of either the RCA or LCA. These anomalies are considered extremely rare and benign. Group II anomalies begin at the normal origin of the right or left coronaries and then take an aberrant course across the base of the heart before returning to their normal routes. Group III is comprised of separately originating LAD and CX arteries from the proximal part of the normal RCA. The single coronary arteries are also given an *R* (right)-type or *L* (left)-type designation, depending on the site of origin, either the left or right sinus of Valsalva. The type III designation, by definition, is always an *R*-type single coronary artery.

Groups II and III single coronary arteries can also be classified based on their relationship to the aorta and PA.¹ The anomalous artery can be anterior, between, or posterior to these vessels. One study¹ modified this classification scheme by adding *septal* when the vessel passes through the interventricular septum and *combined* to describe vessels that follow a combination of routes.

The *R*-I subtype of single coronary artery can be viewed with traditional angiography with either a LAO or RAO view. The *R*-I subtype is characterized by a large RCA that passes through the

AV groove to the anterior base of the heart, giving rise to an anterior descending branch. The *R*-I vessel is responsible for perfusion of the whole heart.

In the *L*-I subtype, the RCA is absent and the CX is markedly dominant and gives origin to the posterior descending branch. The CX then ascends into the right AV groove and provides collaterals to the right atrium (RA) and RV.

The *R*-II and *R*-III anomalies are characterized by origination of the LCA from the right sinus of Valsalva, most commonly from the septum.¹ The *R*-II subtype usually crosses the base of the heart to take up its normal position. The *L*-II subtype, on the other hand, is characterized by RCA origination from the proximal LCA. The most common route it follows is from the LMT, passing between the aorta and PA, to the right AV groove. In the *L*-II subtype, it is rare to find the single coronary artery situated posterior to the aorta or anterior to the PA.¹

ANOMALIES OF INTRINSIC CORONARY ARTERIAL ANATOMY

Congenital Ostial Stenosis or Atresia (LCA, LAD, RCA, CX)

Coronary ostial stenosis or atresia (COSA) is the total absence of an extramural coronary artery. It is characterized by complete absence of the left coronary ostium with a blind ending of the LMT. Blood is thus shunted from the RCA to the LCA, descending anterior artery, and LCX by way of small-caliber

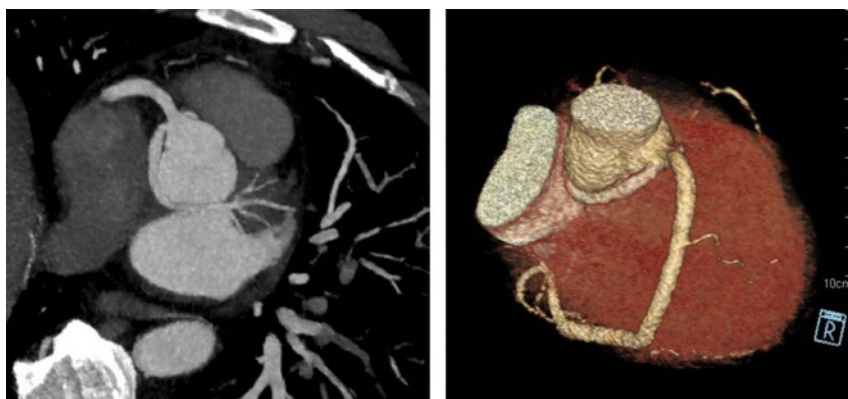


FIGURE 8. Origin of the LMA from the right sinus of Valsalva, retroaortic course; no clinical relevance. Figure 8 can be viewed online in color at www.jcat.org.

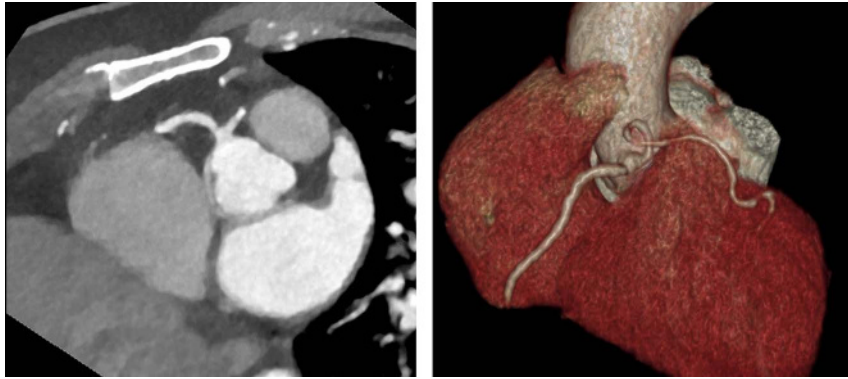


FIGURE 9. Demonstrates a common origin and ostia for the right and left coronary arteries. Figure 9 can be viewed online in color at www.jcat.org.

collaterals. The aortic ostium may be situated ectopically or in its normal position.

The embryology of coronary ostial hypoplasia or atresia is unique in that the coronaries are often normal during early embryonic development but become obstructed or completely occluded later on, in either the fetal or neonatal stage. As such, defects leading to coronary ostial atresia are acquired during later stages of fetal development.⁸

These defects are, by definition, congenital,^{39–44} although evidence suggests that they may also be acquired. Proposed etiologies include atherosclerosis, syphilitic aortitis, arteritis, Takayasu aortitis, or Kawasaki.^{45–47} Ostial stenosis or atresia is often found in association with pulmonary atresia.⁸

Left main coronary ostial stenosis or atresia is more common than right coronary ostial stenosis or atresia. However, both are so rare that they have only been described in case reports. Left main coronary artery ostial stenosis or atresia has been found in association with supravalvular aortic stenosis, right coronary ostium stenosis, and a combination of pulmonary stenosis.^{48–51}

Anomalous coronary artery from the opposite sinus with intramural course is a subtype of COSA. The location of the left ostial sinus of Valsalva varies from that expected based on the location of the distal coronary distribution.⁵² In anomalous coronary artery from the opposite sinus, the proximal trunk is stenotic owing to its intramural location as it passes to the correct side of the heart.

Coronary ostial stenosis or atresia may easily be confused with acquired LMT atherosclerotic occlusion on visualization with CAG.^{53,54} However, the left-sided arteries in hearts with left main COSA are usually smaller in caliber than normal and are not associated with calcifications or atherosclerotic stenosis, thus serving as distinguishing features that can be used to differentiate between the two.⁵⁵ Congenital COSA is characterized by the presence of no more than 2 full-diameter collaterals that do not

narrow at the transition point between the feeding vessel and the receiving vessel. In acquired disease, the collaterals enlarge over time but remain smaller than the receiving vessels. The blind distal LMT of COSA is best appreciated with computed tomographic angiography.¹⁷

Although the RCA is the sole source of coronary circulation in hearts with either a single (right) coronary artery or congenital atresia of the left coronary ostium, they should be considered separate entities. The difference between single coronary artery and COSA is the direction of blood flow. In COSA, flow is occasionally retrograde, from small collaterals to larger left-sided vessels, from the periphery to the center. Collateral flow is supplied by the conal artery and ventricular anastomoses (apical, anterior, posterior, and intraseptal). In a single coronary artery anomaly, flow is always from bigger to smaller arteries, from the center to the periphery.⁵⁶ Whereas coronary ostial atresia is associated with cardiac ischemia, a single coronary artery is not.⁸

Coronary Ostial Dimple

Ostial dimples mark the sites of atretic arteries. Thus, they are often seen as a feature of COSA. The dimples usually consist of fibrous tissue. Ostial dimples are usually followed more distally by proximal coronary stem atresia.¹⁷

Coronary Ectasia or Aneurysm

Coronary ectasia or aneurysm is dilation of an arterial segment to a diameter at least 1.5 times that of the adjacent normal coronary artery. The enlargement may be localized or diffuse. Such dilation may lead to secondary episodic myocardial ischemia. Congenital aneurysms account for 20% to 30% of cases.⁵⁷ Several theories explaining their embryological development have been proposed, such as aneurysm formation secondary to a previous fistula. However, the most widely accepted theory is that

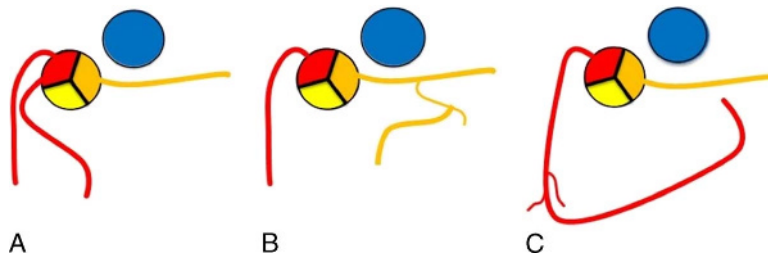


FIGURE 10. Possible anomalies of the CX coronary artery. A, Origin of the CX from the right sinus of Valsalva, retroaortic course; no clinical relevance. B, Origin of the CX from branches of the LAD; no clinical relevance. C, Origin of the CX from distal branches of the RCA; no clinical relevance. Figure 10 can be viewed online in color at www.jcat.org.

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inhibition of the normal coronary vessel development with retention of the primitive sinusoids results in a blind-sac ending. Coronary artery ectasia has been associated with aortic aneurysms.

Acquired cases have been associated with trauma, Takayasu arteritis, polyarteritis nodosa, syphilis, and mucocutaneous lymph node syndrome.⁵⁷ The most common etiology, however, is Kawasaki disease. It has been suggested that the RCA is the most commonly involved. Severe complications, most commonly myocardial infarction and arterial embolization, have been associated with coronary ectasia and aneurysm.^{58–61}

Absent Coronary Artery

As an isolated finding, single coronary artery is a rare anomaly. However, with other congenital anomalies, such as truncus arteriosus and pulmonary atresia, its incidence is much greater. Findings in hearts without LCX arteries include a large dominant RCA that crosses the crux of the heart, ascending through the AV groove to supply both the lateral and posterolateral walls of the heart. In such cases, the LAD is normal in origin and distribution.

Coronary Hypoplasia

Also known as hypoplastic coronary artery disease, this anomaly is characterized by a narrow luminal diameter and shortened course in at least 1 coronary artery or branch. Suggested criteria for diagnosis are luminal diameter less than 1.5 mm and an absence of compensatory branches.⁶² Often, it is present in 2 of the 3 major epicardial branches.⁶² Because of the resulting myocardial hypoperfusion and ischemia, it is associated with sudden unexpected death in otherwise healthy individuals, particularly during exercise. It may be associated with other congenital coronary artery abnormalities, such as anomalous origin of the LCA, left ventricular hypoplasia, or acute angle takeoff of the LCA.^{12,63–66}

Intramural Coronary Artery (Muscular Bridge)

This congenital anomaly is characterized by the partial covering of a coronary artery with myocardial tissue. The most frequently involved coronary is the LAD. Normally, extramural arteries enter the myocardium only once, at their distal end. The abnormal course, however, does not follow this rule and may be sub-epicardial, intussuscepted into the myocardium,^{67,68} intracavitary, or subendocardial.⁶⁹ This can lead to stenosis of the vessel and exercise-induced chest pain. The likelihood of ischemia is positively correlated with the depth of the bridged segment.⁵⁹ The association with sudden death remains unclear; however, it is a consistent cause of myocardial infarction.

With CAG, an intramuscular coronary artery is identified as a squeezing or “milking” effect seen during systole. It may also be seen as a “step down-step up” appearance as the muscular bridge contracts and compresses the artery below, altering coronary blood flow in both a retrograde and anterograde direction.⁷⁰ Muscular bridges may also be visualized using cross-sectional thin maximum intensity projection images on coronary computed tomography angiography (CCTA). Although conventional coronary angiography is the criterion standard for diagnosing myocardial bridges, multidetector computed tomography (MDCT) is also a reliable way to evaluate for myocardial bridging in vivo. The length and depth of muscular bridging can be seen on sagittal multiplanar MDCT reconstruction images.⁷¹ In addition, the vessel can be seen shifting into the myocardium on MDCT.⁷¹

Subendocardial Coronary Course

In rare cases, a coronary artery may pursue a subendocardial course after penetrating the myocardium. This anomaly can be

viewed as an intermediate stage in coronary malposition from intramyocardial to coronary-cameral fistula.

Coronary Crossing

The major branches of the coronary arteries normally course parallel to each other. Coronary crossing is an extremely rare phenomenon, with only 11 cases ever reported in the literature, where at least 2 vessels are found to cross over one another. Reports of involved vessels include the acute marginal branch, diagonal branch, obtuse marginal branch, posterolateral ventricular branch, LAD, LCX, and RCA.⁷² Coronary crossing can be misdiagnosed as ostial occlusion of a normally arising diagonal branch. Normally, coronary arteries do not cross at any point in their individual routes. However, both the origin from the aortic sinus and the routes can be abnormally located and lead to coronary crossing, as seen when the LAD arises posterior to the LCX than normal and crosses it superiorly.⁷² Dynamic cardiac CT can be used to assess the degree of systolic narrowing of the interarterial course.⁷³

Anomalous Origination of the Posterior Descending Artery From the Anterior Descending Branch or Septal Penetrating Branch

The normal vascular supply to the posterior septum is the posterior descending artery (PDA), which arises from the LCX in 10% to 15% of people or RCA in 85% of people.³⁸ Rare instances where the PDA originates from the first septal branch of the anterior descending have been reported. In such cases, the “wrap-around” LAD supplies the descending artery. Left anterior descending continues as PDA across the left ventricular apex.

Split RCA

- Proximal and distal PDAs that both arise from RCA
- Proximal PDA that arises from RCA and distal PDA that arises from LAD
- 2 parallel PDAs (arising from RCA, CX) or codominant

Sometimes referred to as double RCA, split RCA is an anomaly that may have 1 ostium separately arising from the right sinus of Valsalva⁷⁴ or 2 completely separate ostia.⁷⁵ The split RCA can divide from the posterior descending branch, resulting in an anterior subdivision leading to the distal posterior descending branch.⁷⁶ This vessel supplies both the posterior septum and the inferior left ventricular wall as well as the right ventricular anterior free wall. The posterior septum travels within the AV groove, forming the uppermost part of the PDA.⁷⁶ This anomaly is also referred to as *double RCA* on conventional coronary angiography.

Split RCA may easily be missed on conventional coronary angiography, which may explain its seemingly low incidence. On conventional angiography, there may be a question about whether the anomaly is double RCA from a single ostium or a high take-off of a large branch to the RV from the sinus of Valsalva.⁷⁴ However, viewing the vessels from a RAO view may help make the distinction.⁷⁴ Alternatively, CCTA offers 3D views and better accuracy in delineating the origins and courses of the anomalous arteries.⁷⁵

Split LAD

- LAD plus first large septal branch
- LAD, double (parallel LADs)

Normally, the LAD arises from the LCA and courses within the anterior interventricular sulcus (AIVS) to the cardiac apex.



FIGURE 11. Coronary artery fistula connecting between the RCA and LAD artery. There is an obvious aneurysmal process. Figure 11 can be viewed online in color at www.jcat.org.

Dual or split LAD is an uncommon coronary anomaly in which 2 LADs are present. The duplicated LAD may arise as a branch from the LAD proper, the noncoronary sinus, or from the left or right sinus of Valsalva. Often, 1 of the vessels will end well before reaching the apex and thus is referred to as the *short LAD*. The second longer LAD extends to the apex and is called the *long LAD*.⁷⁷

Advances in cardiac imaging have raised the number of known dual LAD subtypes from 4 to 7. Classification is based on the origin and distribution of the long LAD. Both the long and short LAD originate from the LAD proper in types I to III. Types IV to VI are characterized by a short LAD originating from the LMCA and a long LAD originating from the RCA or its sinus. In type VII, both the short and the long LAD originate from the LMCA.⁷⁸ A reported variant of type VII describes the presence of 2 long LADs that extend to the apex in the absence of a short LAD.

In type I, the short LAD originates from the LAD proper and runs within the AIVS, whereas the long LAD runs along the left ventricular side of the sulcus, and then it reenters the AIVS to reach the apex. The left ventricular diagonal branches only arise from the LAD proper or the long LAD. Type II is similar to type I; however, the long LAD descends along the right ventricular side of the AIVS.⁷⁷ Types I and II can be differentiated using a LAO view on CAG. Type III has a short LAD consistent with that seen in types I and II but has a long LAD that travels within the myocardium of the ventricular septum. Type III is best seen in RAO and lateral projections. Type IV dual LAD is the variant in which the longer ectopic LAD arises from the RCA and follows a septal course, and the shorter LAD proper arises from the LCA and terminates high in the anterior interventricular groove.¹³ It supplies septal perforators and diagonal branches.⁷⁹ The short LAD and the LAD proper form a single short vessel located in the superior part of the AIVS.

The type IV long LAD originates from the RCA, whereas the long LAD in type V originates from the right coronary sinus. In both subtypes, the long LAD extends to the mid and distal anterior interventricular groove. The type IV long LAD may travel over the free wall and anterior to the right ventricular infundibulum to reach the anterior interventricular groove or within the myocardium of the septal crest.⁸⁰

The proposed route of a type VI dual LAD is a short LAD from the LMCA giving rise to a large diagonal artery as well as proximal septal perforator arteries. The long LAD of a type VI dual LAD originates from the RCA and travels underneath the right ventricular outflow tract (RVOT) toward the anterior

interventricular groove.⁸⁰ The long LAD gives off small septal perforator arteries throughout its segments.

Ectopic Origination of First Septal Branch

The LAD supplies septal branches to the anterior region of the septum. Ectopic origination of the first septal branch is the result of the persistence of additional, ectopic, or atypical coronary buds that connect with either the RCA or LCA or their branches. Alternatively, they may be associated with anomalous connection of a coronary artery with the opposite sinus or some other structure.

ANOMALIES OF CORONARY TERMINATION

Inadequate Arteriolar/Capillary Ramifications

The small ramifications of the extramural coronaries are terminal. It is probable that an ideal ratio exists with respect to the number of arterioles and capillaries per gram of related myocardium. The ratio could be altered by an anomaly or an acquired condition. It may manifest as angina in patients with otherwise normal coronary arteries, either due to an inadequate number of small vessels or myocardial hypertrophy.

Coronary Artery Fistulas

A coronary artery fistula is a pathologic communication between a coronary artery or 1 of its branches and another vessel (Fig. 11), a cardiac chamber. Although they can be acquired after cardiac procedures or trauma, most are congenital. The majority of documented cases are single coronary fistulas, although rare cases of complex or multiple fistulae have been reported. The most commonly involved artery is the RCA,²² likely because it produces symptoms. The second most-commonly involved artery is the LCA, which is often asymptomatic. Rarely do fistulae with large intracardiac shunts go undetected in children, as they are often associated with signs and symptoms. Fistulae draining into the right side of the heart may cause right ventricular volume overload leading to potentially serious consequences. Depending on their exact locations, fistulas have variable drainage sites including the RV (45%), the RA (25%), and the PA (15%)¹³; however, more than 90% drain into venous circulation.

There are 2 main subtypes of primary coronary arteriovenous fistulas. The first subtype is a fistula between the venous side of the heart and the coronary arterial bed. It usually results in a left

to right shunt. The second subtype is a fistula between the arterial side of the heart and the coronary arterial bed. These patients are usually asymptomatic but on physical examination are found to have a continuous precordial murmur. Chest x-ray often reveals cardiomegaly. Complex coronary fistulae have multiple sites of origin and drainage.

Coronary artery fistulae may also be categorized by size. Small coronary artery fistulae are relatively common anomalies.¹ Most commonly, they arise from a single branch and drain into a single chamber. A study¹ found that the majority originated in the LAD and the drainage point was in the PA. This anomaly rarely causes clinical manifestations and rarely appears to enlarge over time. The study¹ also found that the large fistulae in their patients most often originated from either the RCA or CX arteries. Larger fistulae tend to have tortuous and dilated vessels with the potential to rupture, leading to catastrophic consequences.

The most common type of coronary artery fistula is the coronary artery-cameral fistula. A coronary-cameral fistula is a communication between a coronary artery and a chamber of the heart. Most are congenital and believed to be the result of the persistence of embryonic intratrabecular spaces and sinusoids.⁸¹ The RV is the most frequently involved chamber, followed by the RA, LA, and lastly the LV.

The second most common type of coronary fistula is coronary artery to PA fistula. The most common site of origin is the RCA, followed by the LCA. Because they are usually asymptomatic, most cases are discovered incidentally on imaging, where CT or MR imaging demonstrates draining of the coronary artery or its branches into the PA. Adequate contrast opacification of the coronaries and noncontrast opacification of the pulmonary arteries is required for CCTA demonstration of this fistula type. Their small caliber along with low-pressure circulation and consequent low blood flow may make their evaluation challenging, however.^{82,83}

Coronary artery-coronary sinus fistula is the third most commonly encountered type of coronary artery fistula. On CCTA and MR imaging, the involved arteries appear enlarged and tortuous, whereas the coronary sinus appears dilated. A severely dilated coronary sinus is a hallmark of this type of coronary artery fistula.

A coronary-AV fistula is an abnormal communication between a coronary and a segment of either systemic or pulmonary circulation near the heart that bypasses the capillaries. Coronary artery-superior vena cava (SVC) fistula is another variant documented in the literature. A majority of coronary artery-SVC fistulae drain into a normal SVC. They usually originate from the RCA but may also be found coming from the LCX artery.

Characteristic features of coronary artery fistulae include dilation of the involved vessel, thinned fistulous wall, atherosclerotic changes, thrombosis, fibrosis, and myocardial hypertrophy.⁸⁴ Traditional angiographic imaging of coronary artery fistulae shows retrograde filling of blood out of the arteries.⁸⁵ However, because most fistulae drain into low-pressure chambers, the drainage sites may not be well visualized as dilution of contrast medium often occurs. In addition, traditional angiography does not provide 3D information. An additive value can be achieved, however, when traditional angiography is combined with CCTA to diagnose coronary fistulae. The more frequent use of CT for chest and cardiac imaging has subsequently led to an increase in the diagnosis of coronary artery fistulae as incidental findings. Magnetic resonance imaging and CCTA are the most precise imaging methods for characterization of the fistula anatomy for pretreatment or surgical planning. Imaging is also used after surgery to confirm the complete closure of the fistula.⁸⁶

ANOMALOUS ANASTOMOTIC VESSELS

Extensive interarterial coronary anastomoses (those >40 μm) are observed to occur more frequently in conditions that cause cardiac hypoxia, such as occlusive coronary artery disease, hypertrophy, and valvular disease than in healthy hearts.⁸⁷ Anemia and emphysema are noncardiac conditions also associated with anomalous anastomotic vessels.

CONCLUSIONS

Congenital coronary artery anomalies include a wide array of variants with diverse cardiovascular manifestations. Thorough familiarity with the anatomic pattern and clinical significance of each anomalous condition is necessary to categorize each anomaly and understand its importance. As such, this pictorial review helps radiologists visualize and understand the classification of coronary artery anomalies into those of origin and course, intrinsic anatomy, and termination.

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