Evaluation of anatomy, variation and anomalies of the coronary arteries with coronary computed tomography angiography

Koroner arterlerin anatomi, varyasyon ve anomalilerinin koroner bilgisayarlı tomografi anjiyografi ile değerlendirilmesi

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ABSTRACT

Recent technical advances in computed tomography (CT) have improved image quality, diagnostic performance and accuracy of coronary CT angiography (CCTA). Latest dose-reduction strategies reduce radiation dose to an acceptable level even lower than that from conventional coronary angiography. CCTA is a noninvasive imaging modality which can effectively show complex coronary artery anatomy, variations and congenital anomalies of the coronary arteries. Congenital coronary artery anomalies are rare entities, but sometimes have a potential of producing fatal consequences. CCTA is now the primary imaging modality for the evaluation and diagnosis of coronary artery anomalies. Reporters should, therefore, have knowledge of the normal coronary artery anatomy and variations, and understand the different types of coronary artery anomalies and their respective prognostic implications in order to provide correct diagnosis and to prevent undesirable mistakes during interventional and surgical procedures. *(Anadolu Kardiyol Derg 2013; 13: 154-64)*
Key words: Coronary artery anatomy, variation, anomalies, coronary computed tomography angiography

ÖZET

Bilgisayarlı tomografi (BT) teknolojisinde yaşanan teknik ilerlemeler koroner BT anjiyografinin görüntü kalitesinde, tanısal performansında ve doğruluğunda iyileşmelere yol açmıştır. Yeni geliştirilen doz düşürmeye yönelik stratejiler sayesinde radyasyon dozu kabul edilebilir düzeylere ve hatta konvansiyonel koroner anjiyografi değerlerinin altına inmiştir. Koroner BT anjiyografi invaziv olmayan bir görüntüleme yöntemi olup koroner arterlerin kompleks anatomisini, varyasyonlarını ve konjenital anomalilerini etkili bir şekilde göstermektedir. Konjenital koroner arter anomalileri nadir görülen patolojiler olsa da bazen hayatı tehdit edebilen sonuçlar doğurma potansiyeline sahiptir. Koroner BT anjiyografi günümüzde koroner arter anomalilerinin tanısında primer görüntüleme yöntemi olarak kullanılmaktadır. Bu nedenle raporlayıcıların doğru tanı koyabilmeleri ve cerrahi ve girişimsel işlemler sırasında istenmeyen hataları önleyebilmeleri için koroner arter anatomisini, varyasyonlarını ve koroner arter anomalilerinin farklı tiplerini ve bunların prognostik sonuçlarını bilmeleri gereklidir. *(Anadolu Kardiyol Derg 2013; 13: 154-64)*
Anahtar kelimeler: Koroner arter anatomisi, varyasyon, anomali, koroner bilgisayarlı tomografi anjiyografi

Introduction

Conventional coronary angiography (CCA) is the gold standard imaging modality for the evaluation of coronary artery disease, which is the leading cause of mortality and disability in developed countries. CCA has also a risk of morbidity and mortality, and invasive nature of this expensive technique has led to the development of new imaging modalities that is noninvasive, fast and effective. Recent technical advances in computed tomography (CT) have improved image quality, diagnostic performance and accuracy of coronary CT angiography (CCTA) (1).

The main factors that determine the quality of CCTA are spatial resolution, temporal resolution and volume coverage (2). We need high spatial resolution to visualize the small diameter coro-

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nary arteries and to detect coronary atherosclerosis. Spatial resolution of new generation CT scanners is about 0.4 mm which is inferior from spatial resolution of CCA (0.1 mm). The temporal resolution of current cardiac CT scanners ranges from 135 to 175 ms, and it is possible now to reach a true temporal resolution of 75 ms with dual source imaging. These values are satisfactory to image the heart in diastole without motion artifacts related to heart beating, nevermore very far away from excellent temporal resolution of CCA (4-7ms). New CT scanners have 64-320 detector numbers. An anatomic coverage of 16 cm in one rotation is achieved with 320 detector elements which permit to image the heart in a single rotation within a single heart beat (2).

Radiation dose concerns regarding associated malignancy risk with CCTA have been progressively raised (1). In recent years, the main challenge of CCTA is to reduce radiation dose to an acceptable level lower than that from CCA by the dose-reduction strategies such as tube current modulation, prospectively gated sequential scanning, high-pitch dual source spiral scanning and iterative reconstruction (1, 2). It is now possible to complete a CCTA examination in approximately 1-4 mSv and even lower than 1 mSv (1). For example, in a patient who has heart rates below 65 beats per minute, high-pitch dual source spiral scanning with 256 slice CT allows us to complete a CCTA in a single heart beat with a radiation dose under 1 mSv (2).

Document on appropriate use criteria for CCTA, released in 2010, greatly expands the number of CCTA indications (3). It is very clear that CCTA will gain much a wider application in the field of cardiovascular imaging in the future and supersede CCA as the imaging modality of choice for a broad spectrum of cardiac pathologies (1, 4, 5). Generally, CCTA is the appropriate imaging modality for diagnosis or risk assessment in patients with an otherwise low or intermediate risk of coronary artery disease, and is usually inappropriate for patients at high risk.

CCA is unable to provide sufficient information about the complex anatomy of an anomalous coronary artery because of its distinctive two-dimensional feature, and can detect only 31-55% of anomalies (6, 7). However, CCTA is better than CCA in showing origin, course and termination of the coronary artery, and its relationship to the great arteries and cardiac structures by using multiplanar reformation, three-dimensional volume-rendering and maximum intensity projection techniques (4-6). CCTA sensitivity in visualization of coronary artery anomalies (CAA) is 100% (7, 8). Because of this, CCTA has currently become the method of choice for the diagnosis and preoperative evaluation of CAAs (4, 9). However, training in coronary imaging and familiarity with coronary artery anatomy among radiologists is not satisfactory.

This review article focuses on the normal anatomy, variations and anomalies of the coronary arteries evaluated with CCTA.

Normal coronary artery anatomy and common variations

The aortic root represents the outflow tract from the left ventricle and extends between the aortic valve and the sinotubular junction which demarcates the aortic root from the ascending aorta. Aortic root has three small dilatations called the coronary sinuses (Fig. 1a). More anteriorly located right coronary sinus (RCS) gives rise to the right coronary artery (RCA) and the left main coronary artery (LMCA) arises from more cephaladly located the left coronary sinus (LCS) (Fig. 1b). The posteriorly located aortic sinus normally does not give rise to a coronary artery and named as the noncoronary sinus (10).

Right coronary artery

After originated from the RCS, the RCA runs downwards in the right atrioventricular groove to reach the cardiac crux. The first branch of the RCA is the conus artery in most of the cases which supply the right ventricular outflow tract (Fig. 2a-c). In a small proportion (11.6 to 22%), the conus artery has an origin directly from the aorta (Fig. 3a) which sometimes causes problems during ventriculostomy used to treat ventricular septal defect or pulmonary stenosis (9, 11). The sinoatrial node artery originates from the RCA as the second branch in 55 to 65% of patients (Fig. 2a) and from the proximal left circumflex artery (LCx) in 35 to 45% of the population (Fig. 3a) (10, 12, 13). It can also originate directly from the LCS, RCS, LMCA, (Fig. 3b and 3c) or from the ascending aorta in a small percentage of patients (0.7%) (13). It courses dorsally towards lateromedial aspect of the right atrium. The middle RCA gives rise to several right ventricular branches which supply the anterior free wall of the right ventricle. The largest of them called as the acute marginal branch that runs along the acute margin from base to apex (Fig. 2a). The atrioventricular node artery generally originates from the dominant coronary artery (Fig. 2c) (13). In right coronary dominancy, the RCA gives off the posterior descending artery (PDA) and the posterolateral branch artery (PLB) which perfuse the posterior interventricular septum and the inferior left ventricular wall, respectively (Fig. 2c). The RCA is divided into 3 segments. The proximal segment runs between the ostium and the right ventricular branch. The middle segment is from this point to the acute marginal branch and after, the distal segment courses towards the crux of the heart.

Coronary dominance

Coronary dominance is determined by observing which coronary artery passes over the crux of the heart and gives off the PDA. Mainly, the co-dominance is a state where branches from both the distal RCA and the distal LCx supply the posterior interventricular septum (Fig. 3d) (14). However, some authors define codominance as a state where the PDA originates from the RCA, and the PLB from the LCx (15). Because of these different definitions, the incidence of coronary artery dominance varies in the literature (11). The reported incidence of right dominance is 70% to 89%, left dominance 7% to 13% and co-dominance 2.5% to 20% (11, 13, 15-17). In case of left dominance, the RCA remains in small caliber and terminates before reaching the cardiac crux.

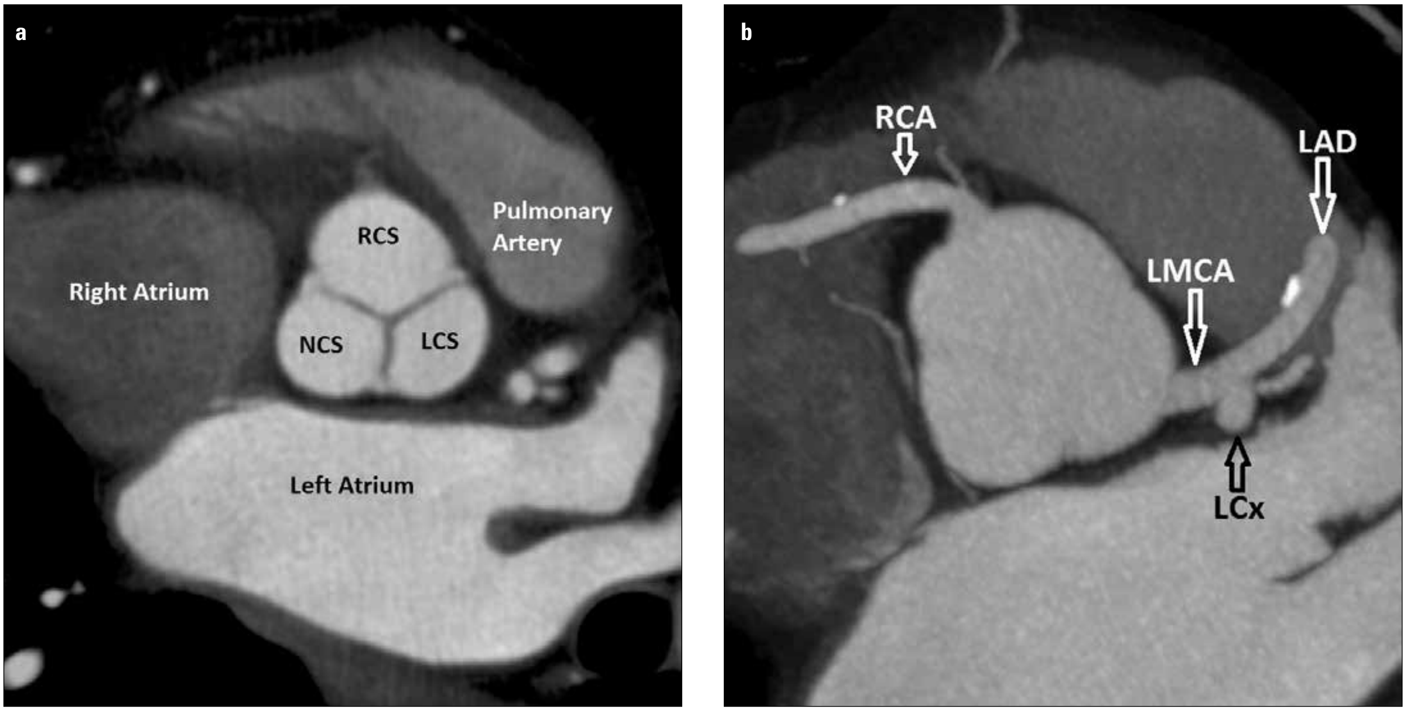

Figure 1. Normal anatomy of the aortic root and coronary arteries. Short- axis coronary computed tomography angiography image (a) shows that the aortic root has 3 aortic sinuses which are the right coronary sinus (RCS), left coronary sinus (LCS) and noncoronary sinus (NCS). Maximum intensity projection reformatted coronary CT angiography image (b) at the level of the coronary sinuses demonstrates that the right coronary artery (RCA) originates from anteriorly located the RCS and the left main coronary artery (LMCA) arises from posteriorly located the LCS. The LMCA divides into the left anterior descending artery (LAD) and the circumflex artery (LCx)

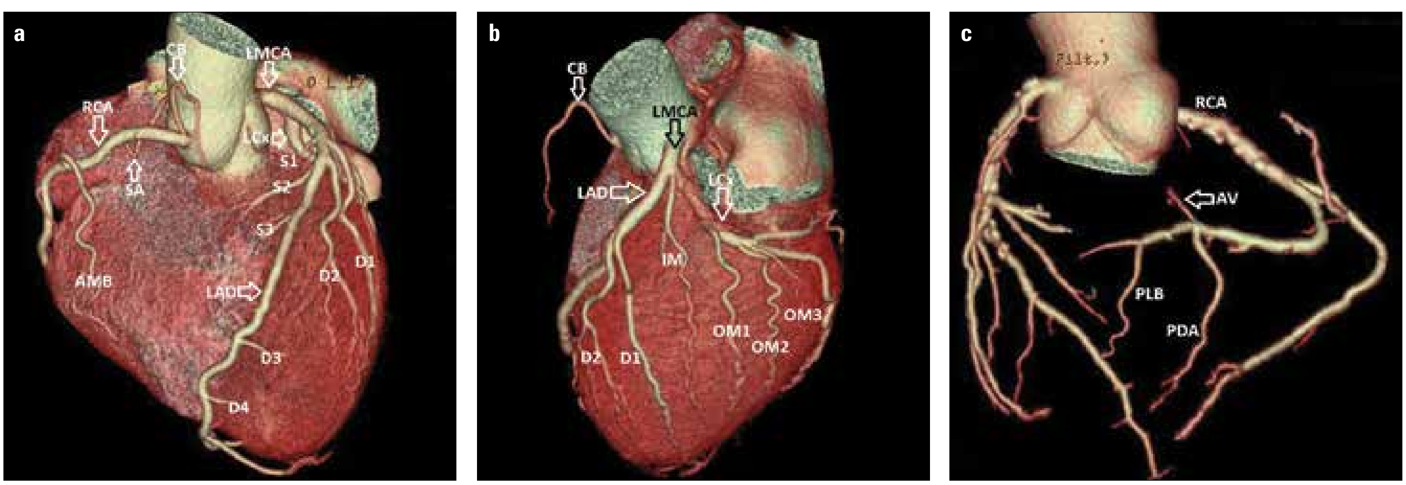

Figure 2. Normal coronary artery anatomy of the heart demonstrated with three-dimensional volume rendered coronary computed tomography angiography images. The conus branch artery (CB) is generally the first branch of the right coronary artery (RCA) and the sinoatrial node branch artery (SA) is the second. The RCA gives also multiple right ventricular branches and the largest of them named as acute marginal branch artery (AMB) (a). The left anterior descending artery (LAD) gives rise to septal (S) and diagonal branches (D), and the left circumflex artery (LCx) gives rise to multiple obtuse marginal arteries (OM) (a and b). The intermediate artery (IM) originates from the LMCA as the third vessel between the LAD and LCx (b). In a right coronary dominancy, the RCA divides into the posterior descending artery (PDA), posterolateral branch artery (PLB) and atrioventricular node artery (AV) at the crux of the heart (c)

Left main coronary artery

The LMCA arises from the aorta, close to the level of the sinotubular junction and slightly cephalad from the origin of the RCA. It than takes a short course between the main pulmonary artery and left atrium. The LMCA divides, beneath the left atrial appendix, into the left anterior descending artery (LAD) and the LCx (Fig. 1b) (9). In approximately 30% of the cases, the LMCA gives rise to the third vessel between the LAD and LCx, called as the intermediate artery (Fig. 2b) analogous to a diagonal branch and usually supplies anterolateral wall of the left ventricle (11, 15). The LMCA length usually varies in between 1-2 cm. Short LMCA (<5 mm) is accepted as a normal variant (Fig. 3b) without clinical significance and found in 4.7% of the cases (11, 13).

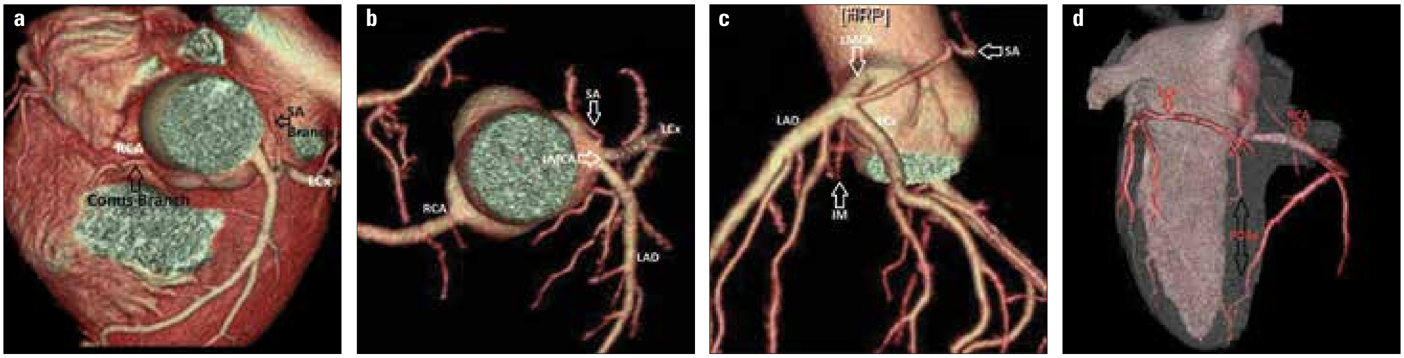

Figure 3. Common variations of the coronary arteries demonstrated with three-dimensional volume rendered coronary computed tomography angiography images. The conus branch artery originates separately from the right coronary sinus and the sinoatrial node branch artery (SA) from the proximal left circumflex artery (LCx) (a). The SA can also originate directly from the left coronary sinus (b) or the left main coronary artery (LMCA) (c) in a small percentage of patients. Also note that the LMCA is short (<5 mm) (b), and tetrafurcates into the left anterior descending artery (LAD), LCX, intermediate artery (IM) and SA (c). Codominancy is a state where the posterior descending artery (PDA) arising from both the distal right coronary artery (RCA) and the distal LCx (arrow) (d)

Left anterior descending artery

The LAD courses downward in the anterior interventricular groove towards the cardiac apex. It gives rise to diagonal branches laterally and septal branches medially which supply to anterolateral free wall of the left ventricle and anterior two thirds of the septum, respectively (Fig. 2a and 2b). The LAD has three segments like the RCA. The proximal and middle segments of LAD is separated from each other by the first septal branch and halfway point from the first septal branch to the apex determines the boundary between the middle and distal segments.

Left circumflex artery

The LCx turns backwards after its origin from the LMCA and runs downwards in the left atrioventricular groove. The proximal portion of the LCx passes under the left atrial auricle that makes the visualization difficult. The LCx gives rise to a variable number of marginal branches and supply the lateral wall of the left ventricle (Fig. 2b). The LCx is divided into proximal and distal portions in relation to the major obtuse marginal branch origin.

Normal diameter of coronary arteries

Diameter of coronary arteries is influenced by gender, body size, coronary dominancy and left ventricular mass, and greatly differs person to person (18). One of the main advantages of CCTA over CCA is its ability to provide visualization of the vessel wall and surrounding soft tissues together with the vessel lumen. So, CCTA can give much more correct measurements about vessel sizes than CCA which gives information only about vessel lumen. Coronary artery lumen diameter and area studies are mainly CCA based and there is need studies with CCTA to define range of normality for coronary artery diameters.

Coronary artery anomalies

The main defect about CAAs is the lack of standardization about classification. In the literature, CAAs are classified either anatomically or clinically (19-21). The most accepted anatomic classification system was proposed by Angelini et al. (19) who divided CAAs into anomalies of origination and course, anomalies of intrinsic coronary arterial anatomy and anomalies of coronary termination. On the other hand, the majority of CAA studies include anomalies in origin and course and sometimes fistulous termination. For example, Yamanaka and Hobbs reviewed 126.595 CCA examinations and only origination and termination anomalies were considered and included as CAA in this large population study (22). Some investigators overrated clinical importance of CAAs and classified them as hemodynamically significant (malignant) and hemodynamically insignificant (benign) (21). Thus, different investigators who used various CAA classification schemes are one of the main factors about different CAA incidence results of CCA studies (20). So, an accepted consensus in the literature about CAA classification system is needed.

The prevalence of CAA is reported to be 0.6-5.6% in patients undergoing CCA, and 0.3% in surgery and autopsy series (20, 22-25). Improved CCTA technology enabled us to detect occult and clinically not important CAAs which otherwise remained undiagnosed by CCA with a high accuracy (4). Thus, after CCTA, detected CAA is increased and ranges between 0.7% to 18.4% (4, 9, 17). The prevalence variability is influenced by the diagnostic method of assessment, entry biases, the criteria used for anomaly classification, and different genetic and geographic backgrounds of the study population (13, 20). They are also found in a higher frequency with congenital heart diseases (20).

Anomalies of origination and course

Absent left main trunk

In case of absent LMCA, the LAD and LCx have separate ostia from the LCS (Fig. 4). This anomaly is observed in a small percentage (0.41%-0.43) of the population, is not hemodynamically significant and is accepted as a variant (13, 22). Sometimes, it may cause technical difficulty in cannulation during CCA and may lead to a faulty diagnosis of occlusion (22). Very short LMCA can be interpreted as absent LMCA with CCA in some cases,

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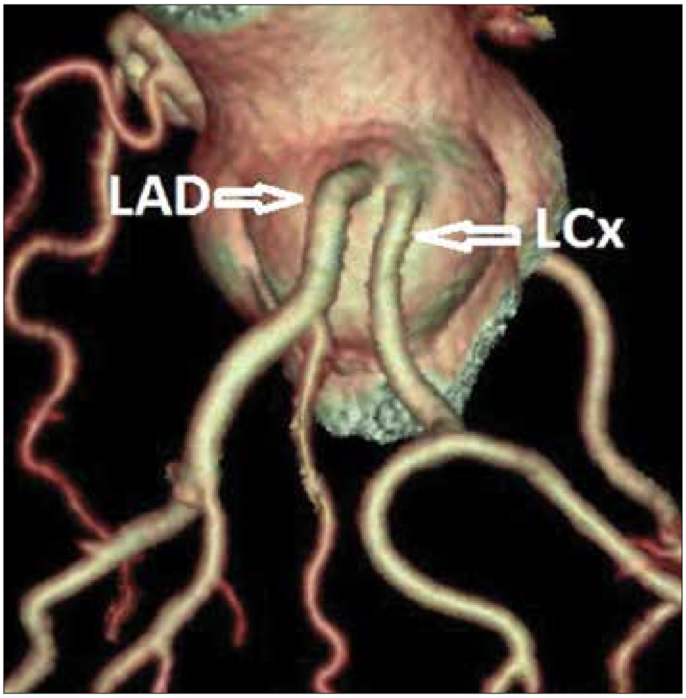

Figure 4. Absent left main coronary artery demonstrated with three-dimensional coronary tree image. The left anterior descending artery (LAD) and left circumflex artery (LCx) have separate ostia arising from the left coronary sinus

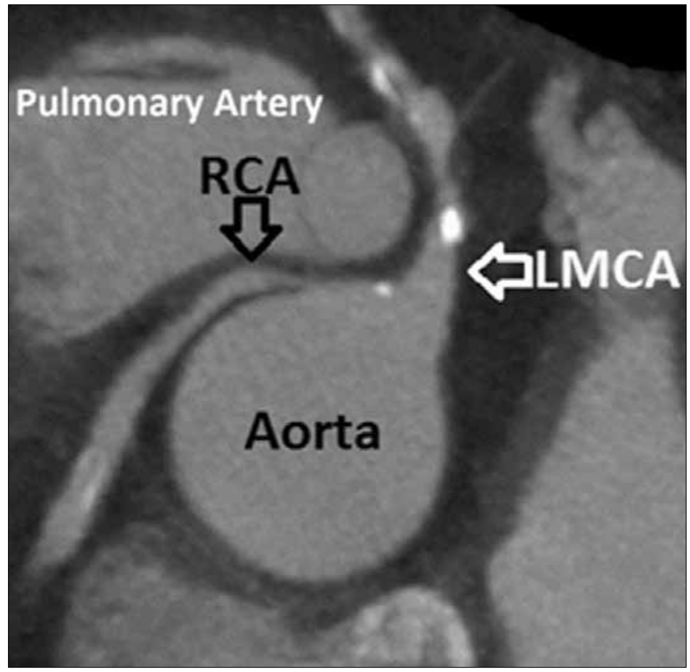

Figure 5. Interarterial course. Axial reformatted coronary computed tomography angiography image shows that the right coronary artery (RCA) originates from the left coronary sinus together with the left main coronary artery (LMCA). Anomalous RCA has an acute angulation and an interarterial course between the aorta and pulmonary artery

and CCTA is superior to CCA in showing the presence or absence of the LMCA (20).

Anomalous location of coronary ostium at improper sinus

A coronary artery arising at improper sinus follows one of the four possible pathways to reach their normal positions. The first and hemodynamically significant pathway is an *interarterial course* (Fig. 5), between the aorta and pulmonary artery, which carries a high risk for myocardial ischemia and appears to be the most common cause of sudden cardiac death in athletic young adults among congenital CAAs (26). Coronary arteries arising at improper sinus and single coronary artery detected with CCA should be clarified with CCTA to illustrate whether an interarterial course exist. The other pathways are accepted as hemodynamically insignificant and these are a *retroaortic course* (Fig. 6a), between the aorta and left atrium; a *prepulmonary course*, anterior to the pulmonary trunk; and a *transseptal course* (Fig. 6b and 6c), through the upper interventricular septum (27). The LMCA originates from the RCS in 0.09-0.2% of patients and the RCA originates from the LCS in 0.03-0.5% of patients (4, 20, 28, 29). Coronary artery origination from a non-coronary sinus is rare. Most cases of the RCA and 75% of the LMCA originating from the opposite coronary sinus takes an interarterial course (22, 28-30). In a study including 27 cases of sudden death in young competitive athletes who have congenital CAA of wrong aortic sinus, it is reported that in 23 cases, the LMCA originated from the RCS and in four cases, the RCA arose from the LCS (26). CAA that most commonly associated with sudden cardiac death is the LMCA originating from the RCS (57%) and the second one is the RCA originating from the LCS (25%) (30). High sudden death prevalence in young ages decreases detected LMCA cases at older ages (4, 31).

Normal coronary ostium has a round or oval shape and an angle 90° or less with the coronary artery (20). Acute angulation of the coronary artery at the improper sinus and its intramural course cause a slit-like appearance to the ostium (20, 26). The true mechanism that induce myocardial infarction leading to sudden cardiac death in young people is not yet completely understood, but impairment of flow by flap-like closure of this slit-like coronary ostium and compression of the anomalous coronary artery between the aorta and pulmonary artery during exercise are proposed to be responsible (17, 20, 26).

The LCx originates from the RCS or the RCA in 0.43% of cases (4). This is a benign anomaly in which almost all the anomalous LCx takes a retroaortic course and has not been associated with death (22, 28). However, retroaortic coursing coronary artery entertains a risk of compression by prosthetic valve fixation rings during valve surgery (4, 22).

Anomalous coronary ostium location

A coronary artery ostium located at the lower end of the coronary sinus is named as low coronary artery ostium and an ostium located within 5 mm of the aortic valve apposition at the aortic annulus is called as commissural coronary artery ostium

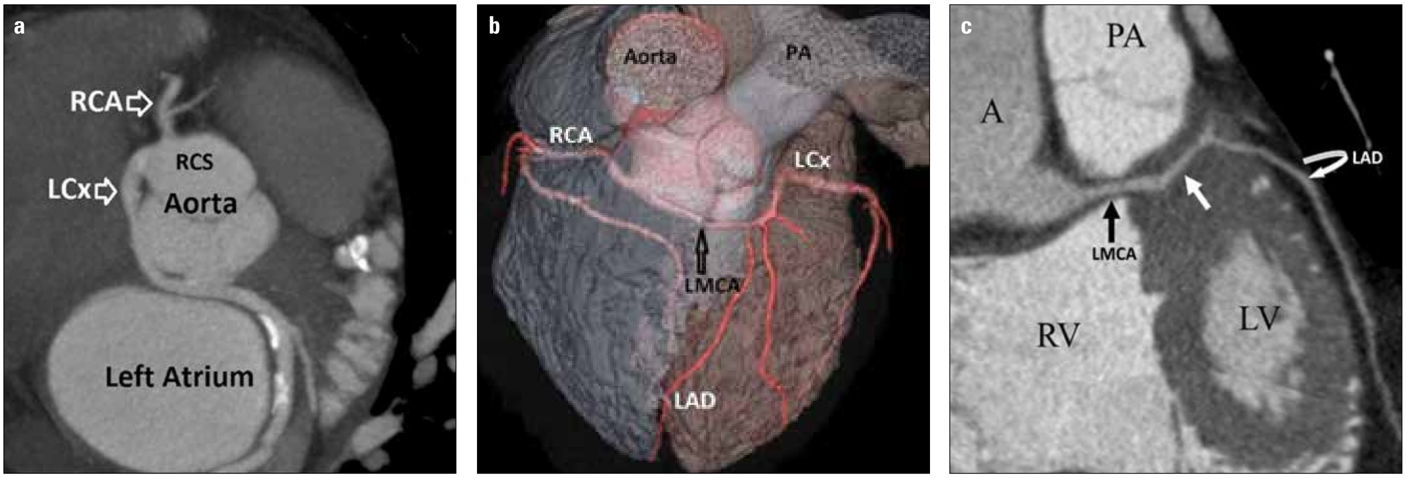

Figure 6. Retroaortic course (a). Curved reformatted maximum intensity projection coronary computed tomography angiography image demonstrates that anomalous left circumflex artery (LCx) arise together with the right coronary artery (RCA) from the right coronary sinus (RCS). The LCx takes a retroaortic course between the aorta and left atrium before reaching the left atrioventricular groove. Transseptal course (b and c). Three-dimensional volume rendered (b) and oblique reformatted (c) images show an anomalous left main coronary artery (LMCA) originating from the RCS and taking a transseptal course within the upper interventricular septum (white arrow)

A - aorta, LV - left ventricle, PA - pulmonary artery, RV - right ventricle

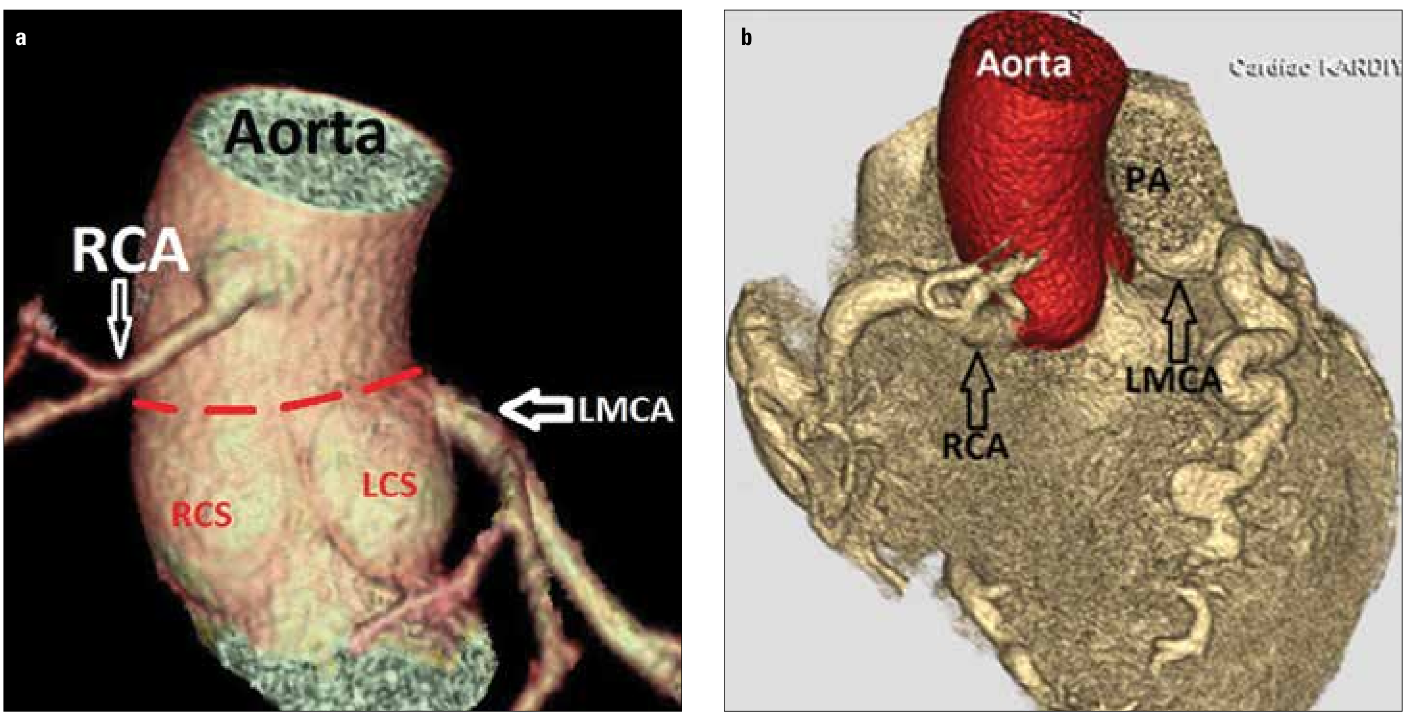

Figure 7. High takeoff. Three-dimensional volume rendered coronary tree image (a) shows that the right coronary artery (RCA) originates from the ascending aorta at least 1 cm above the sinotubular junction (red line) instead of the right coronary sinus (RCS). Note that the left main coronary artery (LMCA) arises normally from the left coronary sinus (LCS). An adult patient with the left coronary artery originating from the pulmonary artery demonstrated by three-dimensional volume rendered images (b). The RCA originates from the aorta and LMCA arises anomalously from the main pulmonary artery (PA). Note that all of the coronary arteries diffusely ectatic and dilated

(20). High takeoff refers to origin of a main coronary artery ostium at least 10 mm above the sinotubular junction (Fig. 7a). It has been reported in 6% of randomly selected adults, but CCTA incidence is under 1% (0.43-0.8%) (4, 11, 20). The RCA is the most common coronary artery that originates above the sinotubular junction (4, 11). This is usually a benign anomaly and carries problem potential during bypass surgery and interventional procedures (22). High takeoff coronary artery can be hemodynamically significant if its orifice is slit-like, has an acute angulation or courses between the aorta and the pulmonary artery resulting in compression and narrowing of the RCA lumen. It is reported that 50% of high takeoff RCA have these findings indicating a relatively high risk of acute coronary attack or sudden death (4, 32).

Anomalous origin of a coronary artery from the pulmonary artery

This is one of the most serious and hemodynamically significant CAA (21). The most common form is the left coronary artery originating from the pulmonary artery, called as ALCAPA syndrome (Fig. 7b) that is seen in 1 of 300.000 live births (33, 34). Infant type of ALCAPA cases is usually symptomatic in infancy and 90% die within the first year of life if untreated (34). Some ALCAPA cases who have sufficient coronary collateral circulation with retrograde perfusion of the left ventricle from the RCA can reach adult life without surgery and with minor cardiovascular symptoms (35). The other form is an anomalous RCA originating from the pulmonary artery (ARCAPA) that is seen much rarer, mostly asymptomatic and is usually detected incidentally in adults (36). CCTA can detect coronary collateral circulation and coronary steal phenomenon indicating a left to right shunt in these cases (4).

Single coronary artery

This rare congenital anomaly is seen in 0.024%-0.066% of patients who undergo CCA and is characterized by a single coronary ostium originating from the aortic sinus (Fig. 8) (20, 22). A single coronary artery is usually asymptomatic, has a benign course and these patients have a normal life expectancy. It can be clinically significant in young people if it has a main branch that course between the aorta and pulmonary artery (37). Single coronary artery also carries a high morbidity and mortality rates in adults in case of critical proximal stenosis before the development of sufficient collateral coronary circulation (37).

Anomalies of intrinsic coronary arterial anatomy

Congenital ostial stenosis or atresia

Congenital atresia or stenosis of the coronary artery is seen rarely and is only reported in sporadic case reports. The LMCA ostial atresia is seen more commonly than the RCA ostial atresia. In case of coronary ostial atresia, the proximal portion ends blindly and blood comes from the other main coronary artery via small collateral pathways such as Vieussens' arterial ring and the Kugel anastomotic artery (38). This coronary anomaly is hemodynamically significant and almost all patients are symptomatic. They usually present with failure to thrive and myocardial infarction in infancy, and syncope in children and adolescents (39). Small caliber collateral circulation is not adequate to the heart and almost all patients need surgical revascularization to prevent the development of myocardial ischemia (38, 39).

Intramural coronary artery (myocardial bridge)

The condition in which a coronary artery segment tunneled within a band of myocardial muscle is named as myocardial bridge (MB) (Fig. 9a). This is accepted to be a benign variant and asymptomatic in most of the cases (13, 22). On the other hand, MB can be clinically significant and may cause various clinical manifestations such as malignant arrhythmias, myocardial

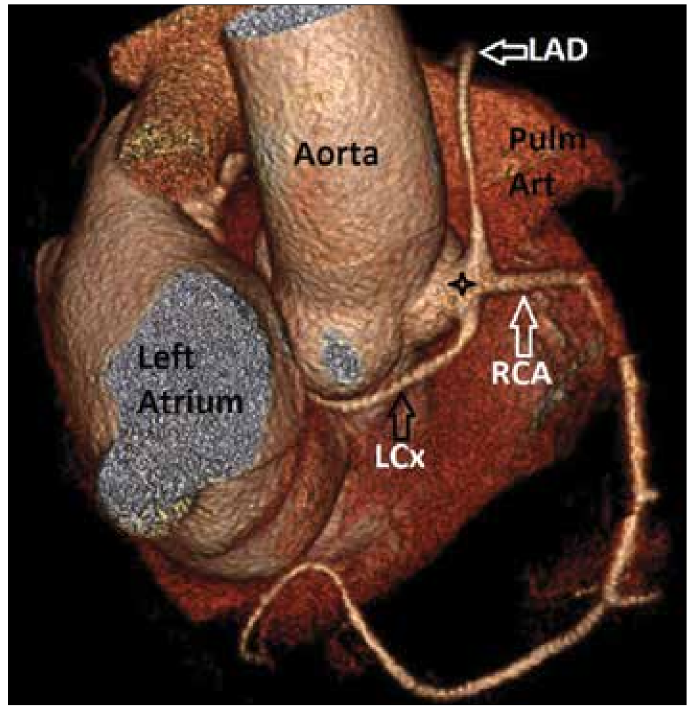

Figure 8. Single coronary artery. Three-dimensional volume rendered posterior view image shows a short single coronary artery (asterisk) originating from the right coronary sinus. The right coronary artery (RCA), left anterior descending artery (LAD) and left circumflex artery (LCx) arise from this single coronary artery and take normal, prepulmonary and retroaortic courses, respectively

infarction and sudden death in rare instances by systolic compression of the tunneled segment persisting into diastole during stress and changing regional hemodynamics that contribute to atherosclerotic plaque formation proximal to the bridge in a higher frequency (40). The likelihood of ischemia also increases with the intramyocardial depth of the tunneled segment (41).

MBs are usually diagnosed incidentally during CCA (20) by the typical findings of 'milking' effect and a 'step down–step up' phenomenon induced by systolic compression of the tunneled segment (40). CCTA gives information about coronary arteries and surrounding myocardium in any plane, and clearly shows the location of the tunneled coronary artery. The great variability of MB between CCTA series (3.5%-58%) and CCA series (0.5%-16%) indicates that CCA is not sensitive enough to detect MB (13, 20, 41). The primary cause of this variability may be the inadequacy of CCA to show superficial MB. On average, MBs are present in about one third of adults and can be seen in any segment of coronary arteries, but most commonly localized in the middle segment of LAD (13, 40).

Split RCA and dual LAD

Split RCA is an extremely rare CAA that is reported only in sporadic case reports. Split RCA appears to be a benign variation. In this anomaly, either there are two RCA originating separately from the RCS or there is one RCA that bifurcates into two major arteries immediately after its origin (42). Split RCA that has two orifices may be overlooked during CCA examinations in case of one orifice cannulation, and again CCTA can accurately show this anomaly.

Dual LAD incidence is reported to be 1% and 1.48% in a CCA and CCTA study, respectively (13, 43). At a rough definition, dual LAD consist of a short LAD which terminates high in the anterior interventricular groove mainly supplying the septal wall, and a long LAD which re-enters the anterior interventricular groove at the distal part, supplying the free wall and the apical wall of the left ventricle. According to the origin and course of the long LAD, dual LAD is classified into four types (43). In the first 3 types, the short and long LADs arise from the proximal LAD (LAD proper). In type 1 (Fig. 9b) and type 2, the long LAD courses on the left ventricular and the right ventricular side of the short LAD, respectively. In type 3, the long LAD has a proximal intramyocardial course within the septum, and appears on the epicardial surface in the distal part of anterior interventricular groove. The long LAD originates from the RCA in type 4.

Split RCA and dual LAD are not important clinically. However, knowing the diseased branch is vital for the cardiac surgeon to prevent an incorrectly placed arteriotomy (11).

Coronary ectasia or aneurysm

Generally, ectasia is used for the whole involvement and aneurysm is used for the partial involvement of the coronary artery when affected vessel diameter exceeds 1.5 times or more the diameter of the adjacent unaffected arterial segment (20). Congenital coronary artery aneurysms are very rare and when detected in pediatric age group, they are usually secondary to Kawasaki disease or coronary artery fistula (44). Coronary ectasia-aneurysm seems to be a variant of atherosclerotic coronary artery disease in the adults and it is very difficult to determine the etiology with diagnostic imaging methods only (13). The prevalence of aneurysmal coronary disease ranges between 1.4% to 4.9% in angiographic, surgical, autopsy and CCTA studies (13, 20). They are found most frequently in the RCA (13). They can be clinically significant and cause thrombosis, rupture, myocardial ischemia, and fistulous communication with neighboring structures (20).

Anomalies of coronary termination

Coronary artery fistula

A coronary artery fistula is an abnormal termination of coronary arteries into either a cardiac chamber, systemic vein, or the pulmonary artery. It has been reported in 0.1-0.2% of patients who undergo CCA and 0.33-0.35% of patients who undergo CCTA (4, 45, 46). It constitutes approximately 15% of CAA (4, 22). The RCA is involved in 44% of cases, the LMCA is involved in 44% of cases, and in 12% of cases, both the LMCA and the RCA are involved (4). The most frequently reported coronary artery fistula drainage sites in CCA studies are the right ventricle (41%), followed by the right atrium (26%) and the pulmonary artery (17%)

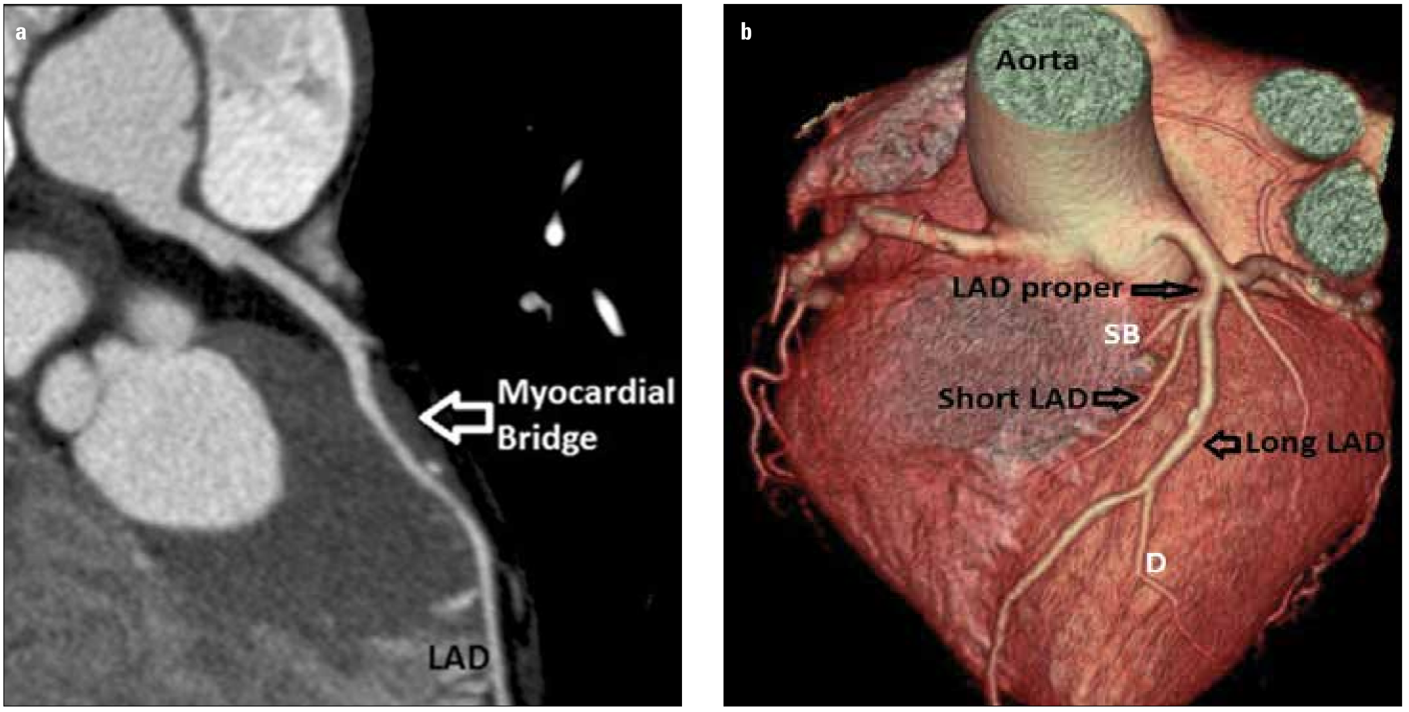

Figure 9. Anomalies of intrinsic coronary arterial anatomy. Typical example of a myocardial bridge (a). Curved reformatted vessel image demonstrates that the middle segment of left anterior descending artery (LAD) has a tunneled, intramuscular course. Three-dimensional volume rendered heart image of dual LAD type 1 (b) shows that LAD proper gives rise to short and long LAD branches. Short LAD terminates in the middle of the anterior interventricular groove and gives rise to septal branches (SB) which supply the upper septum. Long LAD courses on the left ventricular side of the short LAD and re-enters the anterior interventricular groove at the distal part and course to the apex. Long LAD gives rise to a diagonal branch (D) which supply the left ventricular wall

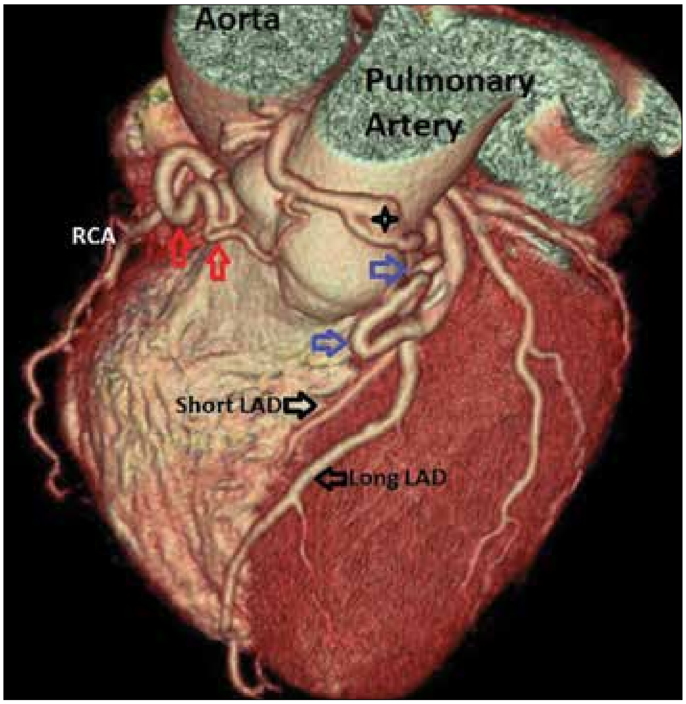

Figure 10. Three-dimensional volume rendered heart image shows a coronary artery to the pulmonary artery fistula. Feeding arteries of the fistula come from both the right coronary artery (RCA) (red arrows) and the left anterior descending artery (LAD) (blue arrows), and the fistula opens to the pulmonary artery via a single orifice (asterisk). This case has also a concomitant dual LAD type 1 (short and long LAD)

(20). In contrast, coronary artery to pulmonary artery fistulas account for 50-89.5% of cases in the CCTA studies (Fig. 10) (4, 46).

Coronary artery fistula drainage sites are clinically more important than the fistula origin, and CCTA is especially superior to CCA in showing fistula drainage site (4). Coronary artery fistulas cause a left-to-right shunt in approximately 90% of cases and have a negative impact on hemodynamic parameters (47). Most coronary artery fistulas are asymptomatic in adults and the severity of the left-to-right shunt determines the clinical presentation (47). High fistula flow causing hemodynamic steal phenomenon, multiple communications, a complex network, multiple terminations and significant aneurysm formations have been associated with a high prevalence of symptoms and complications including myocardial ischemia, congestive heart failure and pulmonary hypertension (4, 20). Early surgical or interventional treatment is recommended because of these important clinical consequences (47, 48).

Hemodynamically significant CAAs

Hemodynamically significant CAA constitutes 20-46% of CAA and carries a risk of malignant arrhythmias, syncope, angina pectoris, myocardial infarction, or sudden death (4, 20, 31). Sudden cardiac deaths in young athletes are caused most frequently by hypertrophic cardiomyopathy (36%) and CAA with its ostium at improper sinus (17%) (49). On the other hand, CAAs are reported to be the most common cause of nontraumatic sudden deaths (61%) in young American military recruits (50). Hemodynamically significant CAAs are an anomalous location of coronary ostium at improper sinus with an interarterial course, an anomalous origin of a coronary artery from the pulmonary artery, a single coronary artery with an interarterial coursing major coronary artery, coronary ostial atresia and a congenital coronary artery fistula (20-22).

Conclusion

CCTA is a noninvasive imaging modality which can effectively show complex coronary artery anatomy, variations and congenital anomalies of coronary arteries. Congenital CAAs are rare and sometimes have a potential of producing fatal consequences. CCTA has gained much a wider application area in the field of cardiovascular imaging after improvement of the radiation dose and is now the primary imaging modality for the evaluation and diagnosis of CAAs. Reporters should, therefore, have knowledge of the normal coronary artery anatomy and variations, and understand the different types of CAAs and their respective prognostic implications in order to provide correct diagnosis and to prevent undesirable mistakes during interventional and surgical procedures.

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References

1. Sun Z, Choo GH, Ng KH. Coronary CT angiography: current status and continuing challenges. Br J Radiol 2012; 85: 495-510. [CrossRef]
2. Hassan A, Nazir SA, Alkadhi H. Technical challenges of coronary CT angiography: today and tomorrow. Eur J Radiol 2011; 79: 161-71. [CrossRef]
3. Taylor AJ, Cerqueira M, Hodgson JM, Mark D, Min J, O'Gara P, et al. ACCF/SCCT/ACR/AHA/ASE/ASNC/NASCI/SCAI/SCMR 2010 appropriate use criteria for cardiac computed tomography. A report of the American College of Cardiology Foundation Appropriate Use Criteria Task Force, the Society of Cardiovascular Computed Tomography, the American College of Radiology, the American Heart Association, the American Society of Echocardiography, the American Society of Nuclear Cardiology, the North American Society for Cardiovascular Imaging, the Society for Cardiovascular Angiography and Interventions, and the Society for Cardiovascular Magnetic Resonance. J Am Coll Cardiol 2010; 56: 1864-94. [CrossRef]
4. Erol C, Şeker M. Coronary artery anomalies: the prevalence of origination, course, and termination anomalies of coronary arteries detected by 64-detector computed tomography coronary angiography. J Comput Assist Tomogr 2011; 35: 618-24. [CrossRef]

5. Kantarcı M, Doğanay S, Karcaaltıncaba M, Karabulut N, Erol MK, Yalçın A, et al. Clinical situations in which coronary CT angiography confers superior diagnostic information compared with coronary angiography. Diagn Interv Radiol 2012; 18: 261-9.
6. van Ooijen PM, Dorgelo J, Zijlstra F, Oudkerk M. Detection, visualization and evaluation of anomalous coronary anatomy on 16-slice multidetector-row CT. Eur Radiol 2004; 14: 2163-71. [CrossRef]
7. Schmitt R, Froehner S, Brunn J, Wagner M, Brunner H, Cherevatyy O, et al. Congenital anomalies of the coronary arteries: imaging with contrast-enhanced, multidetector computed tomography. Eur Radiol 2005; 15: 1110-21. [CrossRef]
8. Kaçmaz F, Özbülbül NI, Alyan O, Maden O, Demir AD, Balbay Y, et al. Imaging of coronary artery anomalies: the role of multidetector computed tomography. Coron Artery Dis 2008; 19: 203-9. [CrossRef]
9. Cademartiri F, La Grutta L, Malagò R, Alberghina F, Meijboom WB, Pugliese F, et al. Prevalence of anatomical variants and coronary anomalies in 543 consecutive patients studied with 64-slice CT coronary angiography. Eur Radiol 2008; 18: 781-91. [CrossRef]
10. Jacobs JE. Computed tomographic evaluation of the normal cardiac anatomy. Radiol Clin North Am 2010; 48: 701-10. [CrossRef]
11. Koşar P, Ergün E, Öztürk C, Koşar U. Anatomic variations and anomalies of the coronary arteries: 64-slice CT angiographic appearance. Diagn Interv Radiol 2009; 15: 275-83.
12. Pannu HK, Flohr TG, Corl FM, Fishman EK. Current concepts in multi-detector row CT evaluation of the coronary arteries: principles, techniques, and anatomy. Radiographics 2003; 23: 111-25. [CrossRef]
13. Erol C, Şeker M. The prevalence of coronary artery variations on coronary computed tomography angiography. Acta Radiol 2012; 53: 278-84. [CrossRef]
14. Schlesinger MI. Relation of anatomic pattern to pathologic condition of the coronary arteries. Arch Pathol 1940; 30: 403-15.
15. Kini S, Bis KG, Weaver L. Normal and variant coronary arterial and venous anatomy on high-resolution CT angiography. AJR Am J Roentgenol 2007; 188: 1665-74. [CrossRef]
16. Eren S, Bayram E, Fil F, Koplay M, Sirvanci M, Duran C, et al. An investigation of the association between coronary artery dominance and coronary artery variations with coronary arterial disease by multidetector computed tomographic coronary angiography. J Comput Assist Tomogr 2008; 32: 929-33. [CrossRef]
17. Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. Circulation 2002; 105: 2449-54. [CrossRef]
18. Dodge JT Jr, Brown BG, Bolson EL, Dodge HT. Lumen diameter of normal human coronary arteries. Influence of age, sex, anatomic variation, and left ventricular hypertrophy or dilation. Circulation 1992; 86: 232-46. [CrossRef]
19. Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation 2007; 115: 1296-305.
20. Sundaram B, Kreml R, Patel S. Imaging of coronary artery anomalies. Radiol Clin North Am 2010; 48: 711-27. [CrossRef]
21. Rigatelli G, Docali G, Rossi P, Bandello A, Rigatelli G. Validation of a clinical-significance-based classification of coronary artery anomalies. Angiology 2005; 56: 25-34. [CrossRef]
22. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary angiography. Cathet Cardiovasc Diagn 1990; 21: 28-40. [CrossRef]
23. Aydar Y, Yazıcı HU, Birdane A, Nasifov M, Nadir A, Ulus T, et al. Gender differences in the types and frequency of coronary artery anomalies. Tohoku J Exp Med 2011; 225: 239-47. [CrossRef]
24. Wilkins CE, Betancourt B, Mathur VS, Massumi A, De Castro CM, Garcia E, et al. Coronary artery anomalies: a review of more than 10.000 patients from the Clayton Cardiovascular Laboratories. Tex Heart Inst J 1988; 15: 166-73.
25. Topaz O, DeMarchena EJ, Perin E, Sommer LS, Mallon SM, Chahine RA. Anomalous coronary arteries: angiographic findings in 80 patients. Int J Cardiol 1992; 34: 129-38. [CrossRef]
26. Basso C, Maron BJ, Corrado D, Thiene G. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. J Am Coll Cardiol 2000; 35: 1493-501. [CrossRef]
27. Ropers D, Gehling G, Pohle K, Maeffert R, Regenfus M, Moshage W, et al. Images in cardiovascular medicine. Anomalous course of the left main or left anterior descending coronary artery originating from the right sinus of Valsalva identification of four common variations by electron beam tomography. Circulation 2002; 105: 42-3. [CrossRef]
28. Kimbiris D, Iskandrian AS, Segal BL, Bemis CE. Anomalous aortic origin of coronary arteries. Circulation 1978; 58: 606-15. [CrossRef]
29. Chaitman BR, Lesperance J, Saltiel J, Bourassa MG. Clinical, angiographic, and hemodynamic findings in patients with anomalous origin of the coronary arteries. Circulation 1976; 53: 122-31. [CrossRef]
30. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. J Am Coll Cardiol 1992; 20: 640-7. [CrossRef]
31. Zhang LJ, Yang GF, Huang W, Zhou CS, Lu GM. Incidence of anomalous origin of coronary artery in 1879 Chinese adults on dual-source CT angiography. Neth Heart J 2010; 18: 466-70. [CrossRef]
32. Wang SP, Jao YT, Han SC. Acute coronary syndrome due to high aortocoronary junction of the right coronary artery: the value of multislice CT. Int J Cardiol 2008; 123: 59-61. [CrossRef]
33. Dodge-Khatami A, Mavroudis C, Backer CL. Anomalous origin of the left coronary artery from the pulmonary artery: collective review of surgical therapy. Ann Thorac Surg 2002; 74: 946-55. [CrossRef]
34. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk: its clinical spectrum, pathology and pathophysiology, based on review of 140 cases with seven further cases. Circulation 1968; 38: 403-25. [CrossRef]
35. Schwerzmann M, Salehian O, Elliot T, Merchant N, Siu SC, Webb GD. Images in cardiovascular medicine. Anomalous origin of the left coronary artery from the main pulmonary artery in adults: coronary collateralization at its best. Circulation 2004; 110: 511-3. [CrossRef]
36. Waite S, Ng T, Afari A, Gohari A, Lowery R. CT diagnosis of isolated anomalous origin of the RCA arising from the main pulmonary artery. J Thorac Imaging 2008; 23: 145-7. [CrossRef]
37. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. Radiology 1979; 130: 39-47.
38. Gebauer R, Cerny S, Vojtovic P, Tax P. Congenital atresia of the left coronary artery--myocardial revascularization in two children. Interact Cardiovasc Thorac Surg 2008; 7: 1174-5. [CrossRef]
39. Musiani A, Cernigliaro C, Sansa M, Maselli D, De Gasperis C. Left main coronary artery atresia: literature review and therapeutical considerations. Eur J Cardiothorac Surg 1997; 11: 505-14. [CrossRef]
40. Möhlenkamp S, Hort W, Ge J, Erbel R. Update on myocardial bridging. Circulation 2002; 106: 2616-22. [CrossRef]

41. Kim PJ, Hur G, Kim SY, Namgung J, Hong SW, Kim YH, et al. Frequency of myocardial bridges and dynamic compression of epicardial coronary arteries: a comparison between computed tomography and invasive coronary angiography. Circulation 2009; 119: 1408-16. [CrossRef]
42. Ökmen AS, Ökmen E. Split right coronary artery: a report of two cases. Turk Kardiyol Dern Ars 2010; 38: 38-40.
43. Spindola-Franco H, Grose R, Solomon N. Dual left anterior descending coronary artery: angiographic description of important variants and surgical implications. Am Heart J 1983; 105: 445-55. [CrossRef]
44. Chakrabarti S, Thomas E, Wright JG, Vettukattil JJ. Congenital coronary artery dilatation. Heart 2003; 89: 595-6. [CrossRef]
45. Vavuranakis M, Bush CA, Boudoulas H. Coronary artery fistulas in adults: incidence, angiographic characteristics, natural history. Cathet Cardiovasc Diagn 1995; 35: 116-20. [CrossRef]
46. Kim MS, Jung JI, Chun HJ. Coronary to pulmonary artery fistula: morphologic features at multidetector CT. Int J Cardiovasc Imaging 2010; 26: 273-80. [CrossRef]
47. Zenooz NA, Habibi R, Mammen L, Finn JP, Gilkeson RC. Coronary artery fistulas: CT findings. Radiographics 2009; 29: 781-9. [CrossRef]
48. Saylan Çevik B, Tavlı V, Sarıtaş T, Oran I, Ergene O. Transcatheter closure of congenital coronary arteriovenous fistula using detachable balloon technique. Anadolu Kardiyol Derg 2010;10: 463-4. [CrossRef]
49. Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. Circulation 2009;119:1085-92. [CrossRef]
50. Eckart RE, Scoville SL, Campbell CL, Shry EA, Stajduhar KC, Potter RN, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. Ann Intern Med 2004; 141: 829-34.