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ORIGINAL ARTICLE

Congenital coronary arteries anomalies: review of the literature and multidetector computed tomography (MDCT)-appearance

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Abstract The prevalence of coronary arteries congenital anomalies is 1 to 2% in the general population. Although the spectrum of their clinical manifestations is very broad from total inocuity to lethal, anomalies of coronary arteries need to be recognized by clinicians in certain circumstances: they are the first cause of death in young adults under physical exercise and an abnormal course of a coronary artery can complicate a cardiac surgery. Therefore, a non-invasive test is highly suitable for detecting anomalies of coronary arteries and multidetector computed tomography (MDCT) is likely to be the best one. To understand how anomalies of coronary arteries may occur, we have reviewed the recent literature about their development. Then, the main types of anomalies are presented with their clinical context, and representative MDCT images from our personal database are used for illustration.

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Introduction

The development of the coronary vasculature has important implications in humans but many variations of their pathway have not been yet fully understood. Nevertheless, knowledge of their development allows the understanding of their variations and anomalies that anatomists and physicians should be aware of. For the last past decades, the diagnosis of coronary arteries anomalies has been based on coronary arteriography. However, Shi et al. [47] recently stated that coronary catheterization failed to take into account of up to 47% of coronary artery anomalies detected using multidetector computed tomography (MDCT). The aim of this review is: (1) to expose a comprehensive view of coronary arteries development and (2) to illustrate MDCT features of major coronary anomalies.

Anatomy

In human, the heart blood supply originates from the ascending aorta—the orifices of the right (RCA) and left (LCA) coronary arteries are located in the corresponding aortic sinus, just distal to the aortic valves. They should be located in the center of each sinus, close to the free edge of the aortic cusp [2]. Both coronary arteries run along the surface of heart in the sub-epicardium and constitute two vascular rings around the heart. The first one runs around the atrioventricular groove and is made of the anastomosis between the RCA and the circumflex artery (Cx) originating from the LCA, while the second one runs along the

interventricular groove and is constituted by the anastomosis between the anterior interventricular branch (AIB), originating from the LCA, and the fourth segment of the RCA, i.e. the posterior interventricular branch (PIB). From the sub-epicardial arteries, small muscular branches are found throughout the myocardium penetrating the ventricular wall, and give rise to arterioles and then to a capillary bed that embraces all cardiac myocytes. The venous return follows muscular and principal arteries and eventually meets in the main collectors: the coronary sinus and the other cardiac veins.

Ontogenesis

Old concepts have suggested that coronary arteries derived from the cardiac mesoderm, like all other cardiac cell types. In the nineties, coronary vessels were thought to derive from the developing aortic sinus wall [48], but the assumption that coronary arteries sprout from the aorta has never been documented by micrographs [49]. Furthermore, this hypothesis failed to explain an origin from the pulmonary trunk. It seems now well established that coronary development is a complex process starting at the surface of the heart and that normally leads to connection with the aorta. Two main processes contribute to the formation of the coronary vasculature—vasculogenesis and angiogenesis. These are followed in the adult heart, in some particular instances by arteriogenesis (i.e. arterialization of pre-existing collaterals, e.g. in ischemic areas).

Development of the heart

Cells that make up the heart (cardiac myocytes, endocardial endothelium cells, vascular smooth muscle cells, vascular endothelium cells and fibroblasts) have different embryonic origins. In human embryo at 3 weeks, the tubular heart is formed as an endothelial tube within a muscular tube, both developed from the lateral plate mesoderm. Cells of the primitive cardiac tube receive their oxygen directly from the ventricular cavity [49] by diffusion through the myocardium. This process is very early and transitory, coronary circulation is essential in the developing embryo. The tubular heart is then submitted to complex morphologic changes including looping, diversification of atria and ventricles, valves formation and septation of the troncus arteriosus. During this period of development, the heart possesses both a myocardium and an endocardium, but lacks an epicardium and blood vessels.

Role of the proepicardial organ

The occurrence of the proepicardial organ (PEO) and its development play a critical role in the apparition of coronary vessels. The coronary vasculature will not develop beyond some primordial stages, in the absence of the PEO. The PEO consists in cells generated from the epithelium derived from the splanchnic mesoderm and associated with the septum transversum [35]. The PEO furnishes an important population of cells to the developing heart, i.e. the epicardium derived cells (EPDC). These EDPC undergo the epithelial to mesenchymal transformation and transform into multipotent cells that cover the heart surface and penetrate the myocardium. The migration of EDPC takes place in four waves that form over the heart three rings, at the level of the atrio-ventricular junction, between the proximal and distal outflow tract, and at the level of the ventriculo-arterial junction. By means of epicardial spurs and islets, the EDPC will cover the whole surface of the heart. This epithelium contributes, as well as the myocardium, to the formation of an extracellular matrix between the epicardium and the underlying myocardium [7].

Development of coronary vessels

As this epithelium thickens, some cells loose contact with its inner wall and migrate into the extracellular matrix of the subepicardial connective tissue space. This epithelial to mesenchymal transition begins in the atrioventricular and the outflow tract grooves [22]. The resulting mesenchyme contributes to the formation of smooth muscle cells, vasculogenic endothelium, pericytes and fibroblasts. These mesenchymal cells massively migrate throughout the developing myocardium [54]. Some coalesce to form pipes within the connective tissue of the atrioventricular groove, and constitute thus the endothelium of the future coronary vessels, whereas others differentiate into arterial smooth muscle cells. These pipes fuse to form a network from which all coronary vessels derive. The way these vessels differentiate either into arteries or veins remains unclear. A primordial non-differentiated vascular plexus makes first contact with the sinus venosus. The blood shows a to-and-fro movement during this stage. The vessels continue to develop into an extensive capillary plexus that spreads along the posterior interventricular groove into the atrioventricular and the anterior interventricular grooves, and then around the bulbus cordis and the base of the truncus arteriosus. Then, this network gives small subepicardial arteries that extend toward the apex, at the surface of the heart, and invade the myocardial wall [6]. This subepithelial plexus is present at Carnegies stage 15–16 (33–37 days gestation) [49]. At the end of the vasculogenic period that occurs in the absence of blood flow, the general pattern of the coronary system is established, but remodeling of the major vessels and capillary bed will take place after connection to the aorta. At this point, no blood flows inside

coronary vessels, so the caliber of coronary vessels depends on other factors than blood flow. However, hematopoietic precursors can be identified within the formed vessels. Endothelial plexi are seen over the surface of the heart and throughout the myocardial wall, but are still not connected to the aorta. The future arterial end of coronary arteries actually grows towards and around the truncus arteriosus. As this latter undergoes septation to form the aortic and pulmonary outflow tracts, vascular channels arising from the peritruncal capillary plexus bypass the pulmonary trunk and selectively contact the proximal aorta, penetrating the tunica media and then the intima at the center of the aortic valve leaflets [31, 51]. For reasons still not understood, the aorta only is invaded and receives connections from the forming coronary arteries [40]. The final connection to aorta occurs during Carnegie stage 18 (44 days of gestation), and the LCA connects earlier than the RCA [49]. Initially, connections are observed to the left, right, and more rarely posterior aortic sinuses. Connection to aorta involves flow-related and apoptotic events that lead to the continuity of coronary endothelium and lumen with that of the aorta [38, 51]. Small coronary vessels in formation probably penetrate the aorta before these connections are functional, providing intermittent blood flow in the system [55]. Some of the penetrating vessels of this endothelial network acquire an adventitia and a layer of vascular smooth muscle cells during the vasculogenesis period [20]. They persist as the final coronary arteries [51], while vessels that fail to develop a tunica media regress [39]. The coronary arteries are thus not outgrowths of the aorta but ingrowths. A possible double origin of the endothelial cells of the most proximal part of the coronary arteries may, however, explain anomalies such as the abnormal origin of the left coronary artery from the pulmonary trunk. Then, coronary vessels are largely remodeled during the development leading to a large panel of variation in adults. At the same period, connection between the venous end of the coronary system and the right atria occurs, forming the coronary sinus and veins [53]. This connection occurs few days before arterial connection in most species [6, 53]. The way the venous end of the coronary system connects to the right atrium remains unclear [53]. Direct shunts between the coronary arteries and veins represent a normal finding in the developing heart and they disappear as the differentiation of the early vessels into arteries and respectively, veins occurs. Persistence of some shunts is not considered an anomaly, in the fully developed heart.

Thus, coronary vessel development is a complex form of vasculogenesis followed by angiogenesis, potentially leading to numerous variations with major implications for clinical settings [3]. Other extracardiac cell lineages also contribute to the formation of the heart, as for example, the neural crest cells [40]—formation of the media of pulmonary trunk and aorta, of the aorto-pulmonary septum, the leaflets of the arterial valves, the parasympathetic cardiac ganglia, the innervation of the coronary arteries, some contribution to the formation of the inflow tracts, and probably also some contribution to the formation of smooth muscle cells of the proximal coronary arteries and the differentiation of the central part of the conduction tissue.

Epidemiology and clinical features

According to a recent paper, congenital anomalies of the coronary arteries occur in 1–2% of the general population [24]. This prevalence is probably underestimated because some anomalies may induce difficulties in coronary artery catheterization. Both genders are equally affected, and no hereditary or epidemiological factor has been observed [30]. Congenital anomalies are usually compatible with normal prenatal heart development and postnatal growth. Usually asymptomatic, these abnormalities may become symptomatic under physical exercise. Symptoms are variable, depending on the type of anomaly and on the age of the subject [3], and include atypical chest pain, dyspnea, cardiomyopathy, ventricular fibrillation, syncope, acute myocardial infarction. For example, the abnormal pathway of a main coronary artery between the aorta and the pulmonary trunk is the second most common etiology of sudden death in healthy young adults [32]. Congenital anomalies of coronary arteries occur as isolated malformations and, because of the particular development of coronary arteries, are not necessarily associated with other congenital heart diseases. Anyway, this association is possible and concerns mainly transposition of the great arteries, tetralogy of Fallot or single ventricle anomalies [34].

Many classifications of coronary arterial anomalies and variations have been in use. The most accepted and of practical relevance is offered by Dodge-Khatami et al. [14], who have described the following conditions—anomalous pulmonary origins of the coronary arteries, anomalous aortic origins of the coronary arteries, congenital atresia of the left main, coronary arterio-venous fistulas, coronary bridging, coronary aneurysms, and coronary stenosis.

Medical imaging of coronary anomalies

Several methods for the diagnosis of coronary anomalies and their pre-operative assessment can be used. Coronary arteriography has long been considered as the gold standard, but congenital anomalies of coronary arteries (e.g. abnormal number and abnormal location of coronary orifices, coronary artery agenesia) may lead to difficulties in coronary artery catheterization. Furthermore, due to the potentially complex 3D features of these anomalies, arteriography may incompletely delineate the course of coronary

arteries [30]. Echocardiography, that has also demonstrated its ability to evaluate anomalous coronary arteries, is limited, however, to the detection of their orifice and their very first segment. Furthermore, echocardiography results depend also on the anatomy of patients and on the acoustic window. Therefore, alternative methods are required for complete diagnosis of coronary anomalies and possibly associated cardiac malformations. Amongst morphological techniques, MRI and MDCT are theoretically able to assess coronary arteries and heart morphology. MRI will probably be the ideal tool in the next few years due to the lack of radiations. At present, however, in clinical practice, its spatial resolution is insufficient to allow a complete study of coronary arteries. MDCT is a non-invasive, non-operator dependent technique that provides high spatial and temporal resolution images of the heart and coronary vessels [27, 47]. Despite irradiation, which can be as low as 5–5.4 mSv when using dose saving algorithms [23], a figure comparable to dose delivered during an invasive coronary arteriography, MDCT is a more-and-more used method for coronary artery assessment.

To illustrate MDCT findings in congenital coronary anomalies, we have selected images from over 700 MDCT cardiac examinations (Sensation 16, Siemens, Forchheim, Germany) performed in our center over a period of 40 months, from September 2003 to December 2006. Acquisitions were performed in supine position with 25–100 mL intra-venous contrast media administration and retrospective electrocardiogram gating. General anesthetic was obtained in children under 6 years. Data were acquired with 16 × 0.75 mm collimation, a 420 ms gantry rotation time, an 80–120 kV tube voltage, and a 50–550 mAseff effective tube current-time product. Axial slices of 0.75 mm thickness were reconstructed with a 0.3 mm increment, using a 160–265 mm field-of-view and a medium-smooth reconstruction filter (B35f).

Anomalous pulmonary origins of the coronary arteries (APOC)

This feature is one of the most serious congenital coronary arteries disorders since 90% of children with such a malformation die before their first birthday. APOC may concern any main coronary arteries and results from an abnormal connection of the peritruncal capillary plexus to the pulmonary trunk. Several features can be found—the prevalences of an abnormal origin of the LCA (ALCAPA, Fig. 1) and of the RCA (ARCAPA) are 0.008 and 0.002%, respectively [57]. This difference is thought to be due to the proximity between the normal aortic location of the LCA orifice and the pulmonary trunk. Rarely, the Cx may be involved, and in extremely rare cases both LCA and RCA are concerned [42], a condition not compatible with patient survival except in the presence of an associated ventricular septal defect [17]. The abnormal orifice is located within any of the three pulmonary sinuses [14]. Associated cardiac anomalies are present in one third of ARCAPA [57] and in 5% of ALCAPA [25], and may include atrial or ventricular septal defect, aortic coarctation, tetralogy of Fallot, aortopulmonary window, double outlet right ventricle, and patent ductus arteriosus. Clinical manifestations depend on the type (ARCAPA or ALCAPA) and the dominant coronary circulation (right or left). Usually, ARCAPA have relatively benign manifestations and may rarely lead to sudden death in case of right dominant coronary circulation [57]. In ALCAPA, also known as Bland-White-Garland syndrome, symptoms can appear during infancy or childhood. The oxygen content of blood entering the LCA from the pulmonary trunk is low but sufficient to meet the demand of the myocardium in neonates [57]. When the pulmonary resistances fall, over the first few days, the onset of symptoms depends on the collateralization between the right and the left coronary circulation, ranging from sudden death in case of insufficient collateralization to absence of symptoms when a massive collateralization allows adequate myocardial perfusion. If the collateral circulation between coronary arteries is adequate, patients may survive and develop a left-to-right shunt from the right coronary artery to the pulmonary trunk [43]. In adults, this steal phenomenon may induce underperfusion and underoxygenation of the left ventricle leading to myocardial ischemia or infarction, congestive heart failure, or cyanosis. Thus, Wesselhoeft distinguished clinical manifestations of ALCAPA according to the ageing of the pathology as follows: (1) infantile syndrome with acute episodes of respiratory insufficiency and sweating, (2) mitral regurgitation with congestive heart failure, (3) syndrome of the continuous murmur, and (4) sudden death in adolescents or adults [56]. Surgical repair must be considered in all cases as soon as the diagnostic is established. ALCAPA must undoubtedly be repaired because of life-threatening outcomes. Surgery in ARCAPA is more debatable but recommended [57]. Surgical options include ligation or aortic reimplantation, bypass graft or Takeuchi operation (intrapulmonary tunnel) [14].

Anomalous aortic origins of the coronary arteries (AAOC)

The AAOC represent one third of all coronary anomalies and regroup abnormal origins of a major coronary artery (i.e. RCA, LCA, Cx or AIB) from the opposite aortic sinus or from the non-facing sinus, abnormal origins of the AIB or the Cx from the RCA, single coronary arteries, inverted coronary arteries, and other unspecified abnormal aortic

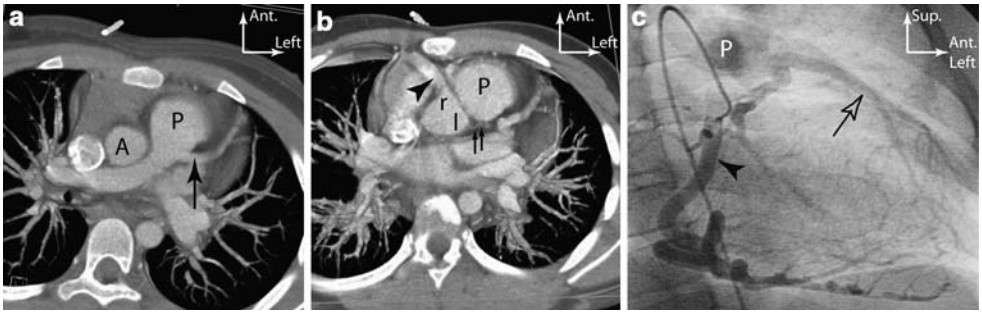

Fig. 1 Volume rendering technique (VRT) 3D-reconstructions from CT images in axial view **a** and **b**, and right coronary arteriography, **c** Bland-White-Garland syndrome in a 13-year old boy with breathlessness. Echocardiography showed a dilatation of the RCA and a reverse flow in the LCA. The LCA (*arrow*) arises from the pulmonary trunk (P); a dilated RCA (*arrowhead*) arises from the right aortic sinus (r); steal phenomenon into the pulmonary trunk via the AIB (*empty arrow*). Note on **b** the small coronary vessel (*short arrows*) arising from the left aortic sinus (l), which may be either an agenesic LCA or an infundibular branch. The LCA has been reimplanted in the ascending aorta. Clinical symptoms resolved after surgery

origins [14]. They usually are considered as benign, except aberrant origin of the LCA from the right aortic sinus and aberrant origin of the RCA from the left aortic sinus. They result either from an abnormal connection to aorta or from an abnormal regression of normally declining vessels.

Origin of a main coronary artery from the non-facing sinus or the opposite aortic sinus

Four patterns have been reported, the RCA and the LCA arising from the left aortic sinus (0.03–0.17%) [58], the LCA and the RCA arising from the right aortic sinus (0.1%) [9], the Cx or AIB artery arising from the right aortic sinus (0.67%) [8] and the LCA and / or the RCA arising from the non-facing sinus (Fig. 2). An abnormal origin per se does not have any particular meaning if not accompanied by slit-like orifice, tangential tract, thinning of the coronary arterial wall, longer intramural tract, or abnormal course. Thus, five different courses are possible for the abnormal vessel: inter-arterial course, i.e. between the aorta and the pulmonary trunk (Fig. 3), pre-infundibular, retroaortic, septal, or posterior to the atrio-ventricular valves (Fig. 4) [2, 44]. An inter-arterial course occurs in 75% of patients with a LCA (or one of its main branches, i.e. AIB or Cx) arising from the right aortic sinus and in 90% of patients with a RCA arising from the left aortic sinus [26]. Clinical symptoms or adverse effects may be present, more frequently when an inter-arterial course of the vessel is present, and these anomalies are potentially lethal. To explain clinical outcomes, several pathological mechanisms have been proposed [5, 32], the increase of the vascular tone of the great vessels under physical exercise leading to compression of the coronary artery, the acute angle (<45°) take-off of the abnormal vessel, the elongation of the intramural tract by the distending aortic or pulmonary root, a long intramural initial segment, closure of the abnormal slit-like coronary orifice, and coronary spasm. The pre-infundibular route can also become a cause of ischemia in case of higher pulmonary pressures with dilatation of the right cardiac chambers and pulmonary trunk. Reported symptoms are disturbance of cardiac rhythm, angina under exercise, syncope or sudden death [32]. Surgery is mandatory and surgical options are reimplantation, bypass graft of the abnormal coronary artery, or unroofing of the intramural coronary segment [5, 14]. In other cases, patients must limit their physical exercise. Lastly, a pre-infundibular course of a major coronary artery may result in special difficulties for heart surgery, as well as a retro-aortic course of the Cx when a mitral annuloplasty or replacement is needed.

Single coronary artery (SCA)

In this abnormality, all coronary arteries arise from a single ostium with a more or less long common trunk (Fig. 5). The prevalence of this abnormality is rare, estimated 0.0024–0.044% [12], associated in 41% with complex congenital heart disease (transposition of great vessels, tetralogy of Fallot, troncus arteriosus, bicuspid aortic valve) [46]. SCA is associated with or without a normal pattern of the coronary distribution, the single artery may divide into two branches that adopted distributions of the right and left coronary arteries, may follow either a right or left coronary artery pattern, or may have a distribution different from the normal coronary tree [29]. Clinical manifestations are absent when no other coronary artery disease or cardiac-associated malformation is present. Potential clinical manifestations are cardiac ischemia, cardiomyopathy, and congestive heart failure. They are mainly due to an abnormal inter-arterial course of one major artery, which may induce sudden death [29]. In these cases, surgical treatment is therefore recommended using coronary artery bypass [16]. Furthermore, rupture of an atherosclerotic plaque of

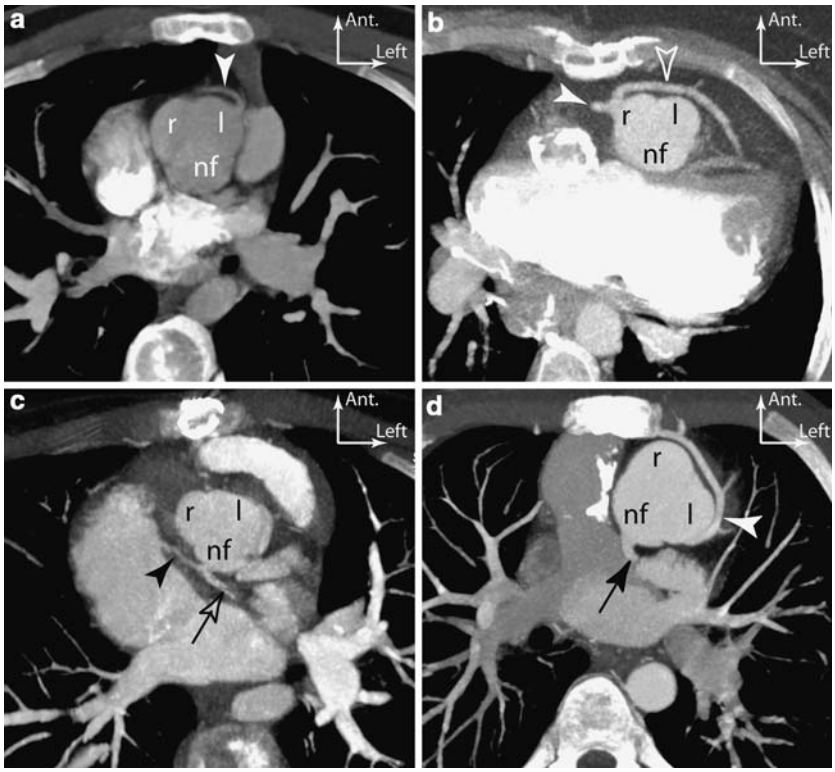

Fig. 2 Maximum intensity projection (MIP) from CT data in 4 different patients—origin of a main coronary artery from the non-facing sinus or the opposite aortic sinus, **a** 18-year-old patient with a corrected tetralogy of Fallot who presented with the onset of ventricular extrasystoles during exercise: the RCA (*arrowhead*) arises from the left aortic sinus (l) and follows a pre-aortic course. **b** A 9-year-old girl with a single ventricle treated by Blalock-Taussig anastomosis and Fontan procedure, suffering from supra-ventricular arrhythmia: the AIB (*empty arrowhead*) arises from the right aortic sinus (r) and follows a pre-aortic course, just posterior to the sternum. Note the common origin with the RCA (*arrowhead*). **c** Follow-up in a 11-year-old boy with corrected double outlet right ventricle, atrial septal defect, and malposition of great vessels, without clinical complaint—the RCA (arrowhead) and the Cx (empty arrow) have a common origin from the non-facing sinus (nf). **d** Follow-up in a 24-year-old man with corrected double outlet right ventricle, malposition of great vessels and ventricular septal defect: the RCA (*arrowhead*) arises from the left aortic sinus (l) and the LCA (*arrow*) from the non-facing sinus (nf)

the common trunk with a proximal stenosis may be devastating because of the inability to develop a collateral circulation [21].

Inverted coronary arteries

In this case, the LCA arises from the right aortic sinus and the RCA from the left one. Then, both arteries have a normal pattern of division [14]. Clinical significance mainly depends on the existence of an inter-arterial course of a major coronary artery, with a risk of sudden death at physical exercise.

Other anomalous aortic origin

Multiple ostia

In their study, Turner et al. [50] found that as many as 35% patients have small accessory coronary orifices. So a separated origin of the RCA and its conus branch from the right aortic sinus (30–50% of normal human hearts) can be considered as a variation, as well as the absence of the LCA associated with a common orifice of both AIB and Cx (Fig. 6a). However, some variations may lead to surgical special problems, for instance, a conus branch arising from the right aortic sinus is at risk during heart surgery, particularly ventriculotomy. The LCA may also be absent with distinct orifices for the AIB and the Cx (0.41%) [11] (Fig. 6b). In few cases, a septal branch may arise directly from the aorta [27]. These conditions provide technical difficulties for coronary artery catheterization but are a source of collaterality in patients with proximal coronary artery stenosis. They can also lead to special difficulties when embolization or alcoholisation of the main septal branch is considered.

High take-off

In this group of abnormal origins, at least one coronary artery rises more than 1 cm distal to the aortic ring, i.e. the

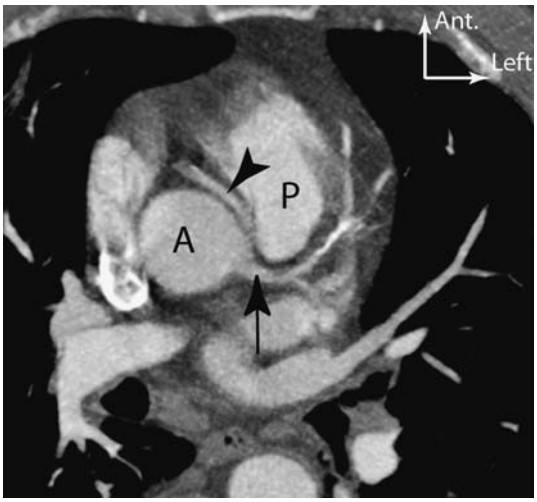

Fig. 3 Axial MIP from CT exam, interarterial course of an abnormal RCA in a 64-year-old man who presented with inferior myocardial infarct, the RCA (*arrowhead*) arises from the LCA (*arrow*) and runs between the aorta (A) and the pulmonary trunk (P). A stent has been implanted on the interarterial segment of the RCA, and angina resolved

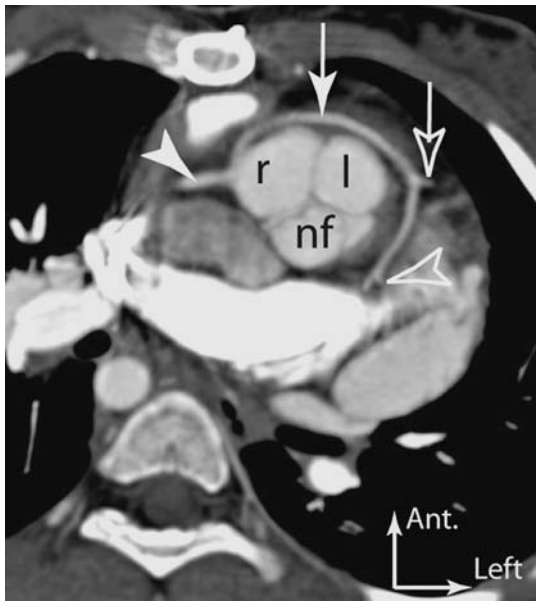

Fig. 5 Axial CT MIP, single coronary orifice in a 10-year-old girl with corrected transposition of the great vessels, ventricular septal defect, and double aortic arch: the RCA (*arrowhead*) and the LCA (*arrow*) arise from the right aortic sinus (r) with a common orifice. The LCA shows a pre-aortic course and gives the AIB (*empty arrow*) and the Cx (*empty arrowhead*). nf: non-facing sinus, l: left aortic sinus

junctional zone between the segment 0 and the segment 1 of the aorta. The prevalence of take-off distal to the aortic ring in the population has been estimated on coronary studies 6% [52]. In almost all cases, they arise from the first centimeter of the ascending aorta, and thus are considered as variations. Abnormally high take-off almost always involve the ascending aorta distal to its first centimeter (Fig. 7), but the aortic arch, the brachiocephalic trunk, the right carotid artery, internal thoracic arteries, bronchial or subclavian arteries and the descending aorta can also be

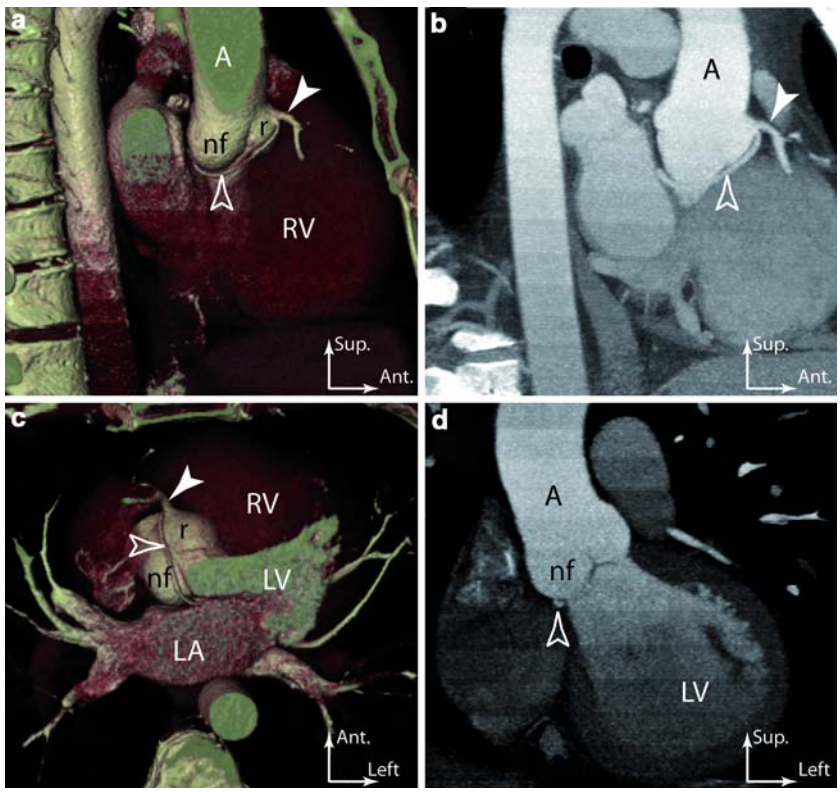

Fig. 4 VRT lateral and caudal views **a**, **c**, sagittal **b** and frontal **d** MIP: abnormal origin of the Cx from the right aortic sinus in a 39-year-old asymptomatic smoker with abnormal ECG-repolarization of the inferior left ventricle wall during physical exercise: the Cx (*empty arrowhead*) arising from the right aortic sinus (r), with a separate orifice from the RCA (*arrowhead*), shows an abnormal retro-aortic course below the non-facing aortic sinus (nf), and reaches the left atrioventricular groove by crossing the mitro-aortic continuity

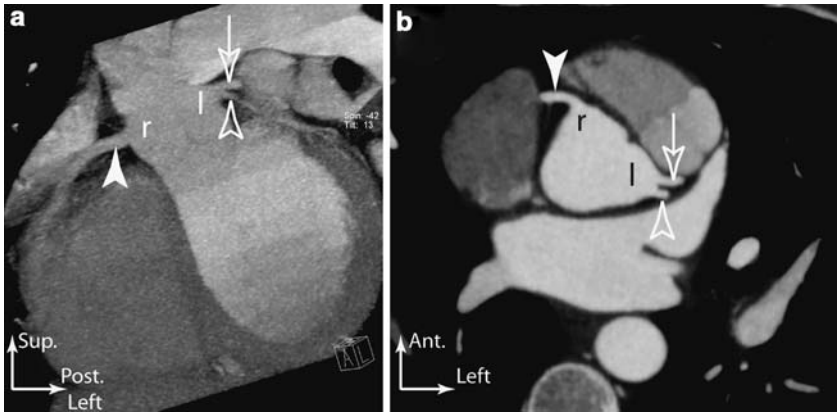

Fig. 6 **a** Short axis CT MIP showing a normal RCA (*arrowhead*) and a common orifice for the Cx (*empty arrowhead*) and the AIB (*empty arrow*), without common trunk of the LCA in an asymptomatic 20-year-old sportsman presenting with an abnormal ECG-repolarization of the inferior wall during physical exercise. The patient was advised against competition. **b** Axial MIP showing three different coronary orifices in a 39-year-old woman referred to emergency unit for syncope, acute coronary syndrome and increased serum troponin level: the RCA (*arrowhead*) arises from the right aortic sinus (r), the AIB (*empty arrow*) and Cx (*empty arrowhead*) arise separately from the left aortic sinus (l)

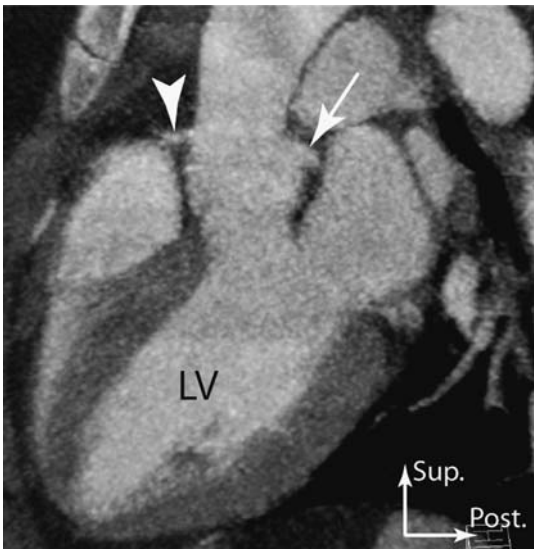

Fig. 7 Sagittal MIP of the aortic root—high take-off of the RCA above the aortic ring in a 12-year-old girl without any other associated cardiac anomaly. She complains about atypical chest pain during exercise: the LCA (*arrow*) originates from the left aortic sinus proximal to the aortic ring; the RCA (*arrowhead*) arises from the right aortic sinus, distal to the aortic ring. No other anomaly was found for explaining symptoms

concerned [2]. These lesions are usually well tolerated and asymptomatic. This abnormal origin may increase the risk of atherosclerotic disease and cause difficulties in canulating the artery during coronarography. According to the distance between the coronary ostium and the aortic ring, the abnormal artery may have a tangential course with respect to the aortic wall and sometimes an intramural course, the coronary artery being embedded in the aortic wall. These patterns may induce coronary flow anomalies leading to clinical symptoms under physical exercise [4]. In these cases, clearly demonstrated using MDCT, surgical reimplantation or unroofing may be proposed. In all cases, cardiac surgeons should be aware of this anomaly in order to cross-clamp and cannulate the aorta high enough to avoid cross-clamping or transaction of the ectopic coronary artery [13].

Congenital atresia of the left main coronary artery (CALM)

In CALM, all three main coronary arteries are located in their normal position but there is no LCA ostium in the aorta (Fig. 8). CALM results from an absence of final aortic connection of the future LCA. The blood feeding to the entire heart depends on the single RCA. The AIB and the Cx flow in a retrograde way, fed by the RCA, and the LCA ends blindly.

CALM has long been considered as a single coronary artery, but the main distinction is that in CALM some of the branches have a reversed blood flow, whereas in a single right coronary artery blood flows anterogradely. CALM is an extremely rare condition, only 33 cases have been reported in the literature until 2003 [15]. In 18% of the reported cases, another congenital heart disease was associated (supravalvular aortic stenosis, ventricular septal defect, and pulmonary stenosis). Because the collateral circulation from the right to the left coronary network is not sufficient to satisfy cardiac metabolism, almost all patients will eventually develop myocardial ischemia, even in the absence of associated coronary atherosclerosis. Clinical findings are almost constant, only 2

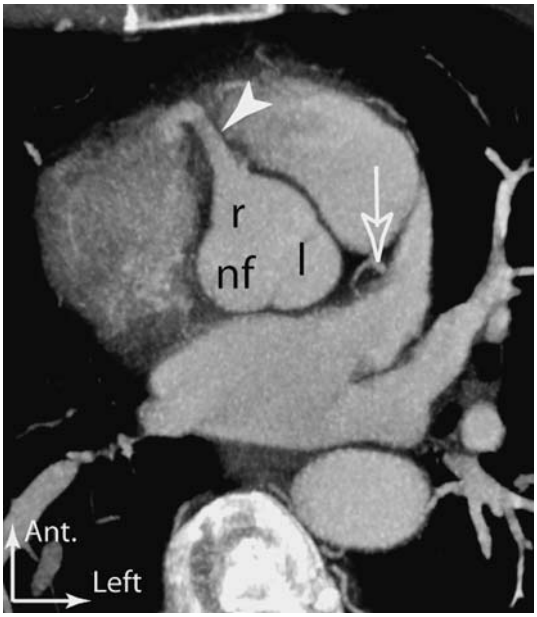

Fig. 8 Axial CT MIP: atresia of the LCA in 53-year-old man who presented typical angina pectoris: dilated RCA (*arrowhead*) arising from the right aortic sinus (r), no LCA was seen from the left aortic sinus (l), the AIB (*arrow*) flowed reversely on right coronary arteriography (not shown) and fed the Cx. Medical treatment was instituted

out of the 33 reported patients were asymptomatic. Symptoms are syncope, myocardial infarction and a delayed growth in pediatrics, and angina and dyspnea in older patients. The prognosis being spontaneously unfavorable, a surgical reconstruction of a two-coronary artery system must be perform using a coronary artery bypass graft [36].

Coronary arteriovenous fistulae (CAVF)

The CAVF is an abnormal communication between a coronary artery and any of the cardiac chambers (Figs. 9 and 10) or the superior vena cava (Fig. 11), the coronary sinus, the pulmonary veins or the pulmonary artery. A case of fistula between the RCA and the left brachiocephalic vein had also been described [37]. The prevalence of CAVF is 0.002% [14]. Their origin is not only congenital, but may also be traumatic, infectious, or iatrogenic [14, 33]. CAVF may be isolated (55–80%) or may occur in association with other congenital heart disease (20–45%: tetralogy of Fallot, atrial septal defect, ventricular septal defect, patent ductus arteriosus, hypoplastic left heart syndrome, and pulmonary atresia with intact ventricular septum) [14, 33]. They involve more commonly the right heart structures and may be multiple in up to 16% [33]. Fistulas originating from both coronary arteries are present in 4–18% of all cases [14]. Sakakibara et al. [45] distinguished two types of CAVF depending on the coronary artery length involved—in type A, only the segment of artery proximal to the fistula is dilated, and the distal end is normal; in type B, the coronary artery is dilated proximal and distal to the CAVF. Clinical symptoms mainly depend on the induced hemodynamic anomalies. CAVF are usually asymptomatic in infants and children, and symptoms occur after 20 years of age—fatigue, angina, dyspnea, or congestive heart failure [33]. All authors agree to close the fistula in symptomatic patients, whereas indication of treatment is controversial in asymptomatic patients. Marvoudis et al. recommend therapeutic intervention in asymptomatic patients when significant clinical, ECG, and roentgenographic findings are present. Therapeutic options include coil embolization or surgical ligation of the fistula with or without coronary artery bypass graft, epicardial ligation in type A or ligation of the pre-capillary end associated with cardiopulmonary bypass in type B [14, 33]. When no treatment is performed, patients must be followed-up. A particular case of coronary fistula is found in pulmonary atresia with intact septum (PAIS). In up to 60% of PAIS, a right ventricle-dependent coronary circulation is present through a non-terminal fistula between the ventricle chamber and either the RCA or the LCA territory [41]. In this particular case, the blood flows from the ventricle to the coronary artery and may supply the left ventricular myocardium. Surgical repair of right ventricle to pulmonary artery continuity, resulting in right ventricular pressure reduction, may induce myocardial underperfusion and ischemia. An alternative approach consists in coil embolization of the fistula prior to surgical repair of the right ventricle outflow tract [18].

Coronary artery bridging (CAB)

The term "bridging" refers to an intramyocardial course of a normally sub-epicardial coronary artery. The prevalence of CAB varies from 5.4 to 85.7% [14]. CAB mainly concerns the middle segment of the AIB (up to 80%), and because of its own prevalence must be considered as a variation of this particular artery rather than an anomaly [2]. CAB can although be located in any other coronary artery [1, 14], and are then abnormal [2]. The classification is based on the involved artery, the presence or absence of symptoms, and the percentage of systolic stenosis during stress test [14]. Symptoms, when present, are angina, myocardial infarction, arrhythmias, or sudden death [1]. Symptoms are related to systolic and diastolic lumen occlusion of the involved segment [19], vasospasm [10], or thrombosis due to mechanical traumatism of the coronary wall. Therapeutic options include β-blocker therapy to decrease heart

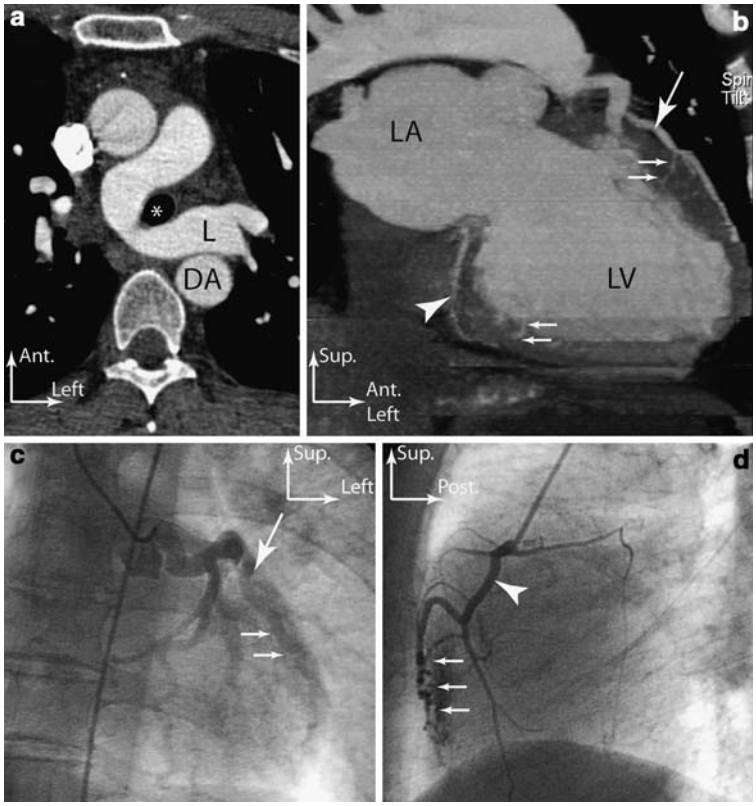

Fig. 9 Axial CT scan at the level of T4 **a**, long axis MIP **b**, left **c** and right **d** coronary arteriography views—diffuse coronary-to-ventricular chamber fistulas in a 36-year-old woman with left ventricle non-compaction and angina pectoris: fistulas between the RCA and the AIB, and the left ventricle chamber are seen through the myocardial wall (*short arrows*). Note on **a** an associated retro-tracheal (*) left branch of the pulmonary trunk (L). DA: descending aorta, LA: left atrium, LV: left ventricle. Patient has been medically treated

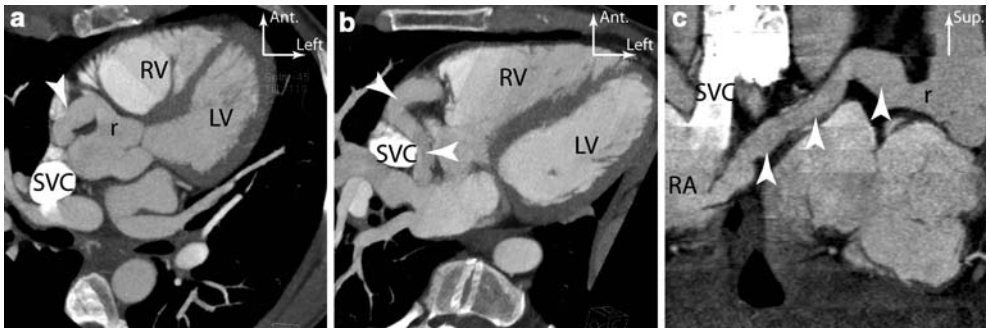

Fig. 10 CT MIP **a** and **b**, and curved MIP **c**—isolated fistula between the RCA and the right atrium in a 41-year-old woman suffering from atypical chest pain: large dilatation of the RCA (arrowhead) that arises from the right aortic sinus (r), and reaches the right atrium (RA) after a tortuous course. SVC, superior vena cava; LV, left ventricle; RV, right ventricle. No surgery was performed

contractility, stent implantation, myotomy, or coronary artery bypass graft [14].

Coronary artery aneurysms (CAA)

The CAA is defined as a dilatation of a coronary segment 1.5 times the adjacent normal segment (Fig. 12). The incidence of CAA is 0.3 to 4.9% [14] with a clear predominance in male (88%). Aneurysms may be congenital or acquired (atherosclerosis, systemic diseases, iatrogenic, infectious and traumatic), fusiform or saccular (with a higher risk of rupture and thrombosis), unique, multiple or diffuse. In congenital CAA, the RCA is the most often involved artery (68%), followed by the LCA (39%), the AIB (17%) and the Cx (17%) [28]. Symptoms are non specific, indicating a coronary artery disease [28]. Surgery is controversial, CAA does not warrant intervention per se and surgical management depends on clinical manifestations, the presence of coexistent coronary artery stenosis, or an enlargement of the CAA [28]. Surgical treatment is based on coronary artery bypass graft, aneurysmorraphy, and/or thrombo-endarterectomy [14, 28].

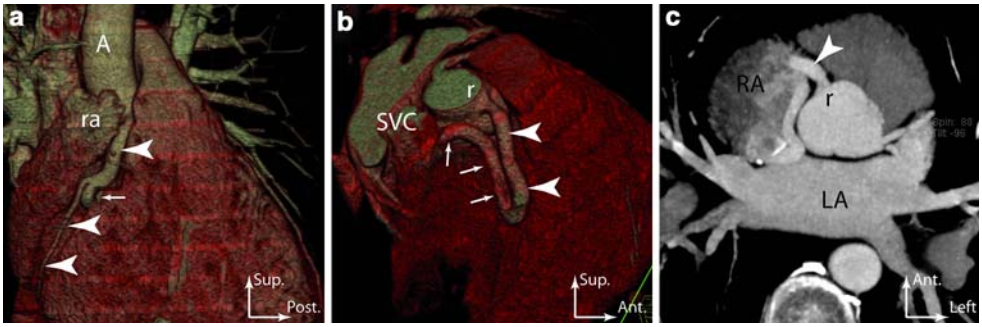

Fig. 11 CT VRT **a** and **b**, and axial MIP **c**—isolated fistula between the RCA and the superior vena cava in a 22-year-old man suffering from a recent onset of angina: large dilatation of the RCA (*arrowhead*) that arises from the right aortic sinus (r), and reaches the superior vena cava (SVC) after a loop. The right appendage (ra) was edited out on a workstation in order to demonstrate the course of the fistula **b**. Note that the second segment of the RCA has a normal diameter in the atrioventricular groove, so the proximal segment of the RCA proximal to the fistula (*short arrows*) is solely enlarged. A surgical ligature was performed, and the patient is free of symptoms at 3-year follow-up. A, aorta; LA, left atrium; RA, right atrium

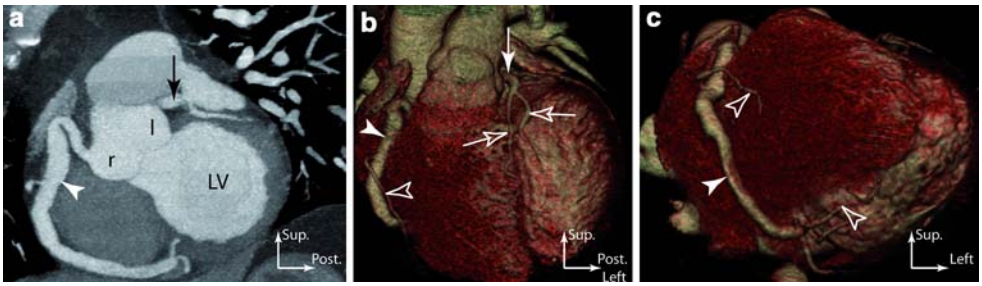

Fig. 12 Short axis MIP **a**, anterolateral **b** and antero-inferior **c** VRT views—coronary aneurisms in a 49-year-old man who presented 2 inferior myocardial infarcts—diffuse fusiform aneurysm of the RCA (*arrowhead*) with normal marginal branch and PIB (*empty arrowheads*), diffuse fusiform aneurysm of the LCA (*arrow*) with local fusiform aneurysm of the AIB and of the first diagonal branch (*empty arrows*). Anticoagulant treatment was instituted. LV, left ventricle; r, right aortic sinus; l, left aortic sinus

Coronary stenosis (CS)

Coronary stenosis are almost always acquired, but congenital CS have although been described [48]. Stenosis are classified either in ostial or peripheral, and according to the coronary segment involved [14]. Coronary orifice stenosis may be due to a valve-like ridge of aortic wall, to hyperelastosis of the aortic wall, or to the fusion of aortic leaflets and aortic wall. Symptoms and therapeutic approach are the same as in atherosclerosis stenosis, except the age of onset.

Conclusion

Coronary artery anomalies are not infrequent, and may be life threatening when an abnormal course of a main coronary artery exists. Physicians and surgeons should suspect these anomalies when an atypical chest pain or ECG-abnormal repolarization occurs in a young adult or a child, particularly under physical exercise. MDCT is the first rank diagnosis tool to perform when an abnormal origin of coronary arteries is suspected or needs to be ruled out.

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