7.8 Evaluation of the Coronary Anomaly, Fistula, Aneurysm, and Dissection

S. Fröhner

Contents

7.8.1 Introduction

Non-atherosclerotic lesions of the coronary arteries present as a manifold and heterogeneous spectrum with different clinical relevance and distinguishing congenital and acquired forms. The acquired non-atherosclerotic diseases of the coronary arteries have different causes. They can develop over a long time and remain clinically inconspicuous, or they can result in life-endangering acute clinical symptoms. Catheter angiography is not able to demonstrate all of these anomalies in detail, especially complex anomalous courses, because selective probing and subsequent interpretation of vessel anatomy is difficult and the angiographer is not aware of an atypical location of the vessel orifice.

Multi-slice CT, in particular the newly available 16- and 64-slice CT scanners with their ability to generate nearly isotropic voxels, represent a highly promising tool to visualize coronary arteries with respect to their origin and course. MPR, thin MIP, SSD, and VRT allow for accurate visualization of the coronary arteries, including the detection and characterization of anomalies of the coronary artery system.

7.8.2 Anomalies of the Coronary Arteries

Coronary artery anomalies constitute 1–3% of all congenital malformations of the heart. In approximately 0.46–1% of the normal population, anomalies of the coronary arteries are found incidentally during catheter angiography or autopsy.

The etiology of coronary artery anomalies is still uncertain. Maternal transmission of some types has been suggested, particularly when only a single coronary artery is involved. Familial clustering is also reported for one of the most common anomalies, in which the left circumflex coronary artery (CX) originates from the right sinus of Valsalva. Anomalies of the coronary arteries may also be associated with Klinefelter's syndrome and trisomy 18 (i.e., Edwards syndrome). Cardiac causes for early and sudden infant death include anomalies of the coronary arteries; the Bland-White-Garland-Syndrome may be one relevant cause. Anomalies of the coronary arteries found in children may be associated with other congenital anomalies of the heart like Fallot's syndrome, transposition of the great arteries, Taussig-Bing heart (double-outlet right ventricle), or common arterial trunk.

The normal anatomy (Fig. 7.47) of the coronary arteries is well-known. The left main coronary artery (LMA) arises from the left sinus of Valsalva (LSV) and bifurcates into the left anterior descending (LAD) and CX. The right coronary artery (RCA) originates from the right sinus of Valsalva (RSV).

Anomalies of the coronary arteries can concern their origin, course, and aberrant distal branches.

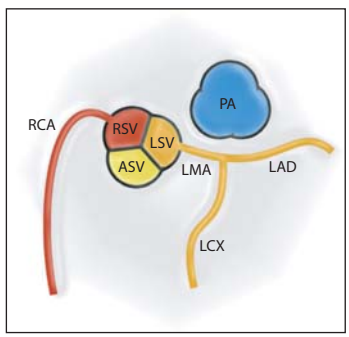


Fig. 7.47. The normal anatomy of the coronary arteries

Common variants are anomalies with origin from the contralateral side of the aortic bulb. These include an origin of the LMA or the LAD from the RSV or the proximal RCA and an origin of the RCA from the LSV or the LMA. There are four possible pathways for these aberrant vessels to cross over to their regular peripheral locations: (1) "anterior course" ventral to the pulmonary trunk or the right ventricular outflow tract, (2) "interarterial course" between the pulmonary artery and aorta, (3) "septal course" through the interventricular septum, and (4)"retro-aortic course". Clinically, course anomalies of the coronary arteries are subdivided into "malignant" and "non-malignant" forms. Malignant forms are associated with an increased risk of myocardial ischemia or sudden death and mostly show a course between the pulmonary artery and aorta (i.e., "interarterial"). The most common case is an origin of the RCA from the LSV that courses between the aortic bulb and the pulmonary artery. Anomalies of the LMA or the LAD arising from the RSV with a similar course are associated with higher cardiac risk, too. It is suggested that myocardial ischemia and sudden death result from transient occlusion of the aberrant coronary artery, due to an increase of blood flow through the aorta and pulmonary artery during exercise or stress. The reason is either a kink at the sharp leftward or rightward bend at the vessel's ostium or a pinch-cock mechanism between the aorta and pulmonary artery. Up to 30% of such patients are at risk for sudden death. Courses of anomalously long coronary arteries ventral to the pulmonary artery may be associated with a higher risk of myocardial ischemia. Dilatation of the pulmonary trunk in pulmonary hypertension can lead to a stretching mechanism of the aberrant vessel that causes lumen reduction and consecutive myocardial ischemia.

An origin of the left (or right) coronary artery from the pulmonary artery (Bland-White-Garland-Syndrome for the left coronary artery) has to be considered as malignant. The LMA arises from the left or posterior aspect of the pulmonary trunk immediately above a pulmonary sinus of Valsalva. If collaterals are well-developed, the RCA and its major branches are dilated and tortuous. This anomaly is frequently associated with myocardial ischemia and sudden death in early childhood. Clinically silent forms can also be found. The aberrant coronary artery is mostly fed by collaterals from the other coronary arteries. Early detection is mandatory for successful and surgical intervention, which is absolutely required. Other courses do not lead to clinical symptoms, but are incidental findings in catheter angiography, CT of the coronary arteries, or autopsy. In case of planned cardiac surgery, these anomalies nevertheless acquire increasing importance in order to avoid being endangered by accidental surgical injuries, for example, a retro-aortic course of the CX in mitral valve surgery. Therefore, every anomaly of origin and course of the coronary arteries should be described accurately prior to cardiac surgery. If catheter angiography does not provide detailed information, multi-slice CT should be performed to depict the exact courses of aberrant coronary vessels. Finally, aberrant coronary arteries have been discussed to carry a higher risk of arteriosclerosis, in particular a retro-aortic-coursing CX. Thus, careful attention must be paid to these vessels in patients with atypical chest pain or pathological ECG findings.

The following figures are a diagrammatic collection of the most important and frequent coronary anomalies for the LMA (Fig. 7.48), LAD (Fig. 7.49), CX (Fig. 7.50), RCA (Fig. 7.51), and other complex anomalies (Fig. 7.52). The exclamation mark in the figure indicates a malignant anomaly. Coronary anomalies can be reliably identified with 16-slice CT and 64-slice CT, as demonstrated by the case examples given in Figures 7.53–7.57.

7.8.3 Coronary Artery Fistulas

Coronary artery fistulas are a rare condition representing arteriovenous communications between one or several coronary arteries and the right heart chambers (right atrium, right ventricle, 65%) or central veins (superior or inferior vena cava, coronary sinus, or pulmonary artery, 17%). The hemodynamics resemble those of an extracardiac left-to-right shunt.

In many cases, the amount of blood flow through the coronary fistula is small. Other cases represent "high-flow" coronary fistulas with diminished per-

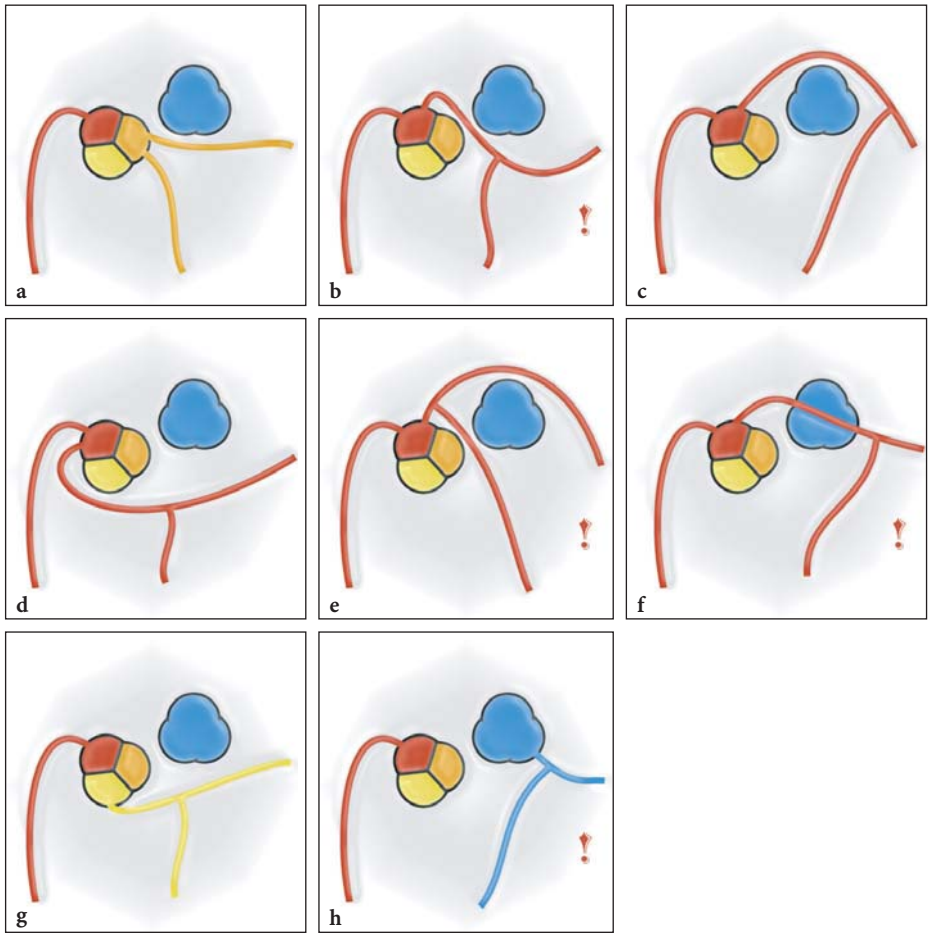


Fig. 7.48a–h. Possible anomalies of the left main coronary artery (LMA). **a** Separate ostia of the LAD and LCx from the left sinus of Valsalva; no clinical relevance. **b** Origin of the LMA from the right sinus of Valsalva, interarterial course; "malignant". **c** Origin of the LMA from the right sinus of Valsalva, anterior course; no clinical relevance. **d** Origin of the LMA from the right sinus of Valsalva, retro-aortic course; no clinical relevance. **e** Origin of the LMA from the right sinus of Valsalva, anterior course of the LAD, interarterial 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 pulmonary artery; "malignant"

fusion of the portion of the myocardium supplied by the arterial feeder of the fistula. A so-called hemodynamic steal phenomenon may occur. In high-flow fistulas, the feeding coronary artery is dilated and often tortuous. Focal saccular aneurysms may develop, which eventually can become calcified and/or thrombosed; aneurysm ruptures have also been reported. Up to 50% of such fistulas arise from the RCA.

The anatomy of coronary artery fistulas can range from abstruse courses up to a spider-like configuration and they can arise from more than one coronary artery. Numerous communications between the coronary artery and the terminating localization can rule out the need for surgical correction. Pediatric patients tend to be especially symptomatic and may present with atypical and typical chest pain and myocardial ischemia associated with ECG alter-

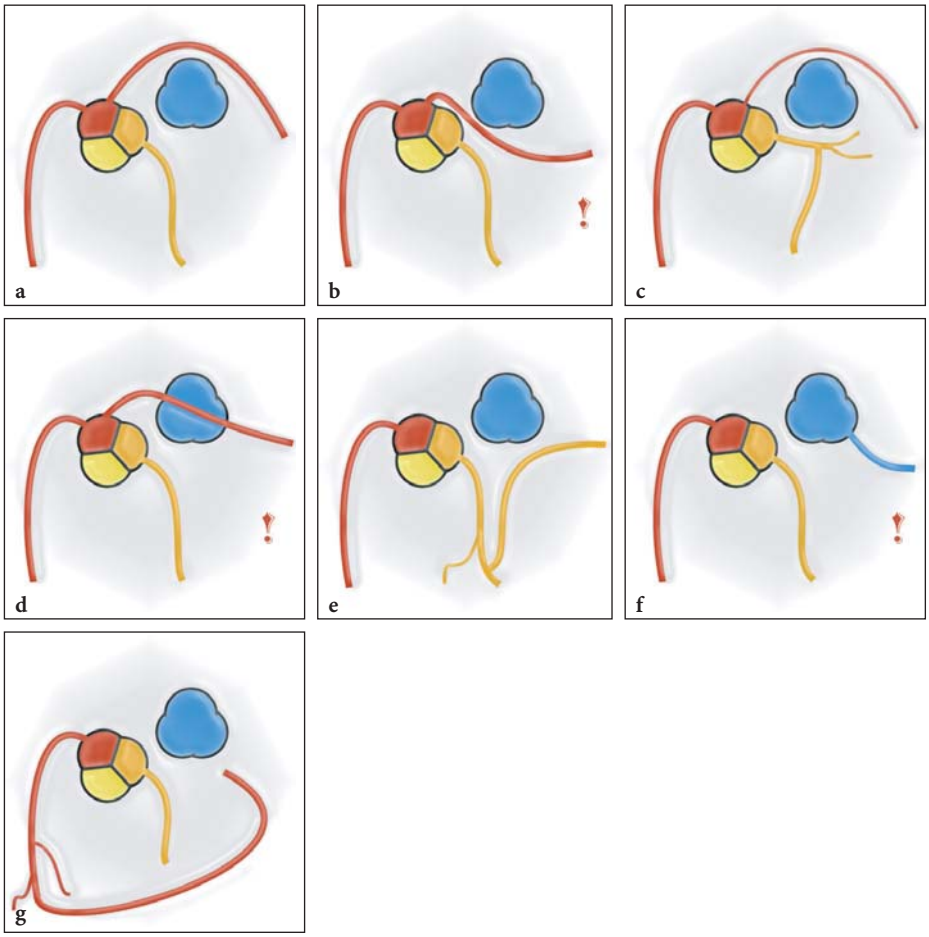


Fig. 7.49a–g. Possible anomalies of the LAD. **a** Origin of the LAD from the right sinus of Valsalva, anterior course; no clinical relevance. **b** Origin of the LAD from the right sinus of Valsalva, interarterial course; "malignant". **c** Origin of distal branches of the LAD from the right sinus of 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 pulmonary artery; "malignant". **g** Origin of the LAD from the distal RCA; no clinical relevance

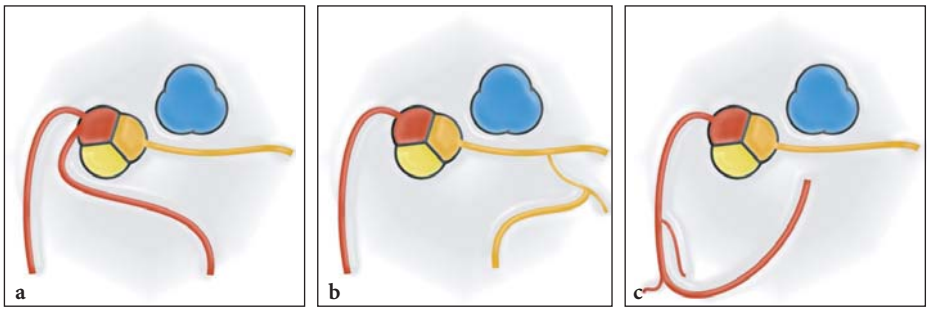


Fig. 7.50a–c. Possible anomalies of the CX. **a** Origin of the CX from the right sinus of Valsalva, retro-aortic course; no clinical relevance. **b** Origin of the CX from branches of the LAD; no clinical relevance. **c** Origin of the CX from distal branches of the RCA; no clinical relevance

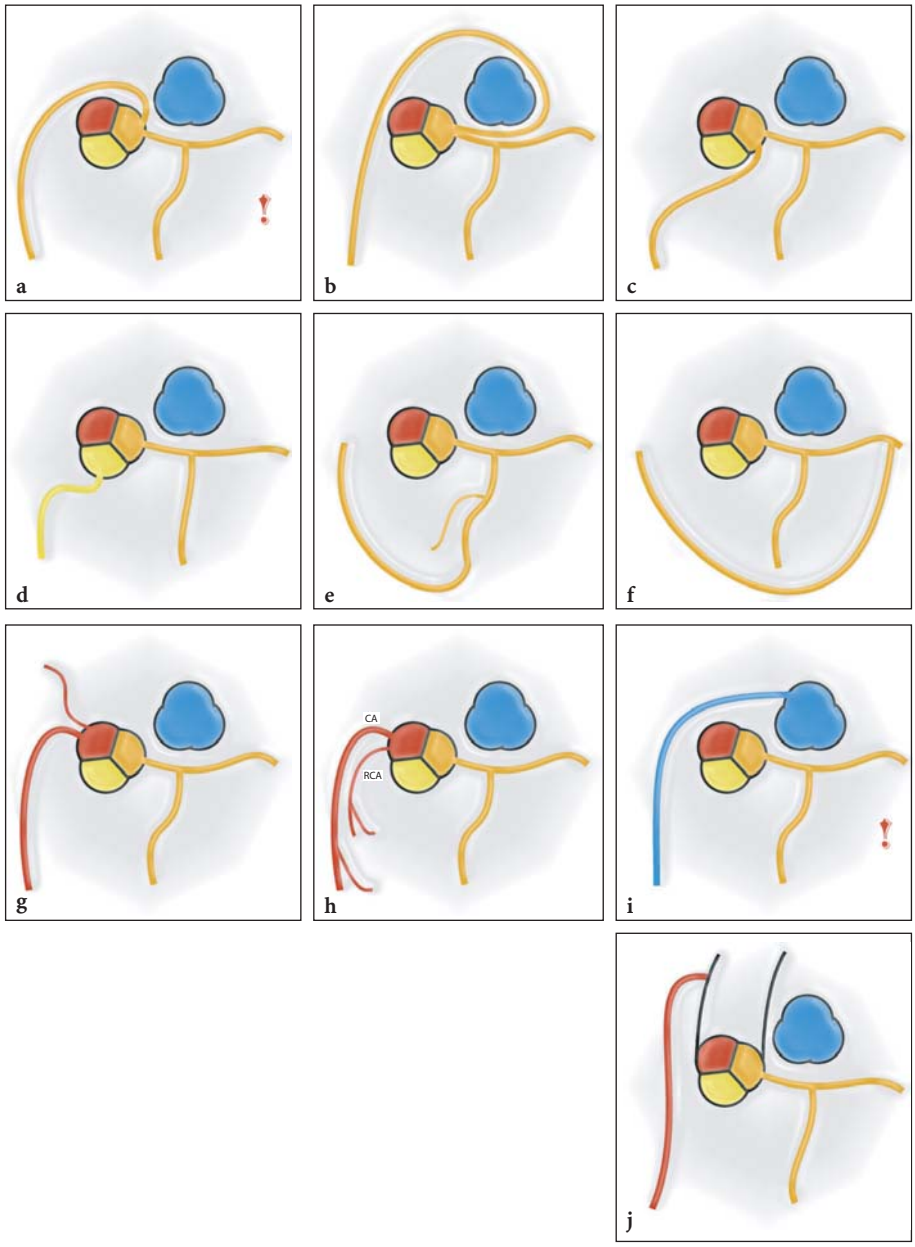


Fig. 7.51a–j. Possible anomalies of the RCA. **a** Origin of the RCA from the left sinus of Valsalva, interarterial course; "malignant". **b** Origin of the RCA from the left sinus of Valsalva, anterior course; no clinical relevance. **c** Origin of the RCA from the left sinus of Valsalva, retro-aortic 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 pulmonary artery, 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

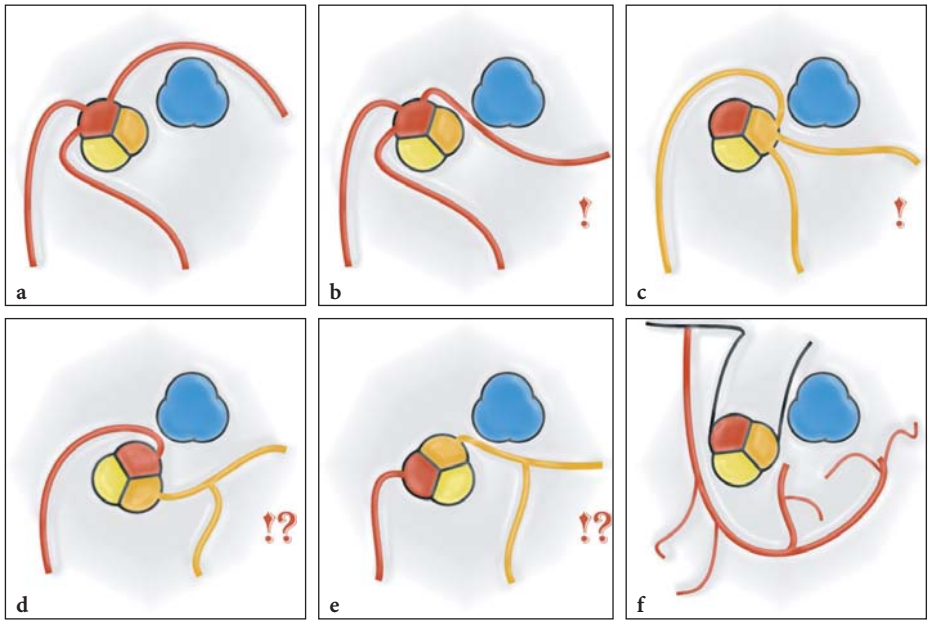


Fig. 7.52a–f. Other complex coronary anomalies. **a** Origin of RCA, CX, and LAD from the right sinus of Valsalva, anterior course of the LAD; no clinical relevance. **b** Origin of RCA, CX, and LAD from the right sinus of Valsalva, interarterial course of the LAD; "malignant". **c** Origin of RCA, CX, and LAD from of 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

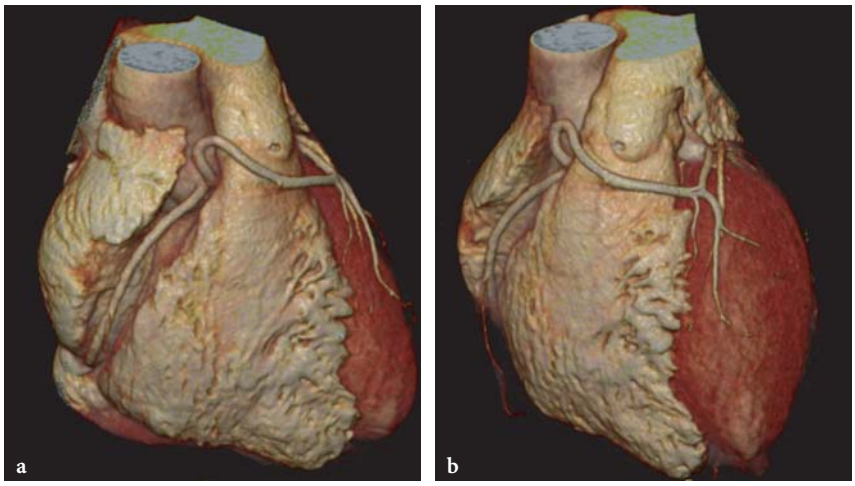


Fig. 7.53a,b. 16-slice CT examination demonstrating the origin of the LMA from a common ostium with the RCA at the right sinus of Valsalva. The artery courses ventral to the pulmonary artery to the left side. Volume-rendering reconstructions in ventral view (**a**) and along the course of the LMA ventral to the pulmonary trunk before the artery branches into the LAD and CX (**b**). Note the ectopic origin of the first diagonal branch out of the LMA

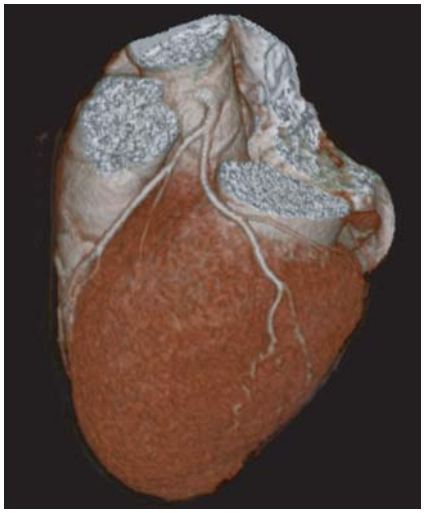


Fig. 7.54. 16-slice CT examination showing an ectopic origin of the LMA cranial to the left sinus of Valsalva; displayed with volume-rendering reconstruction

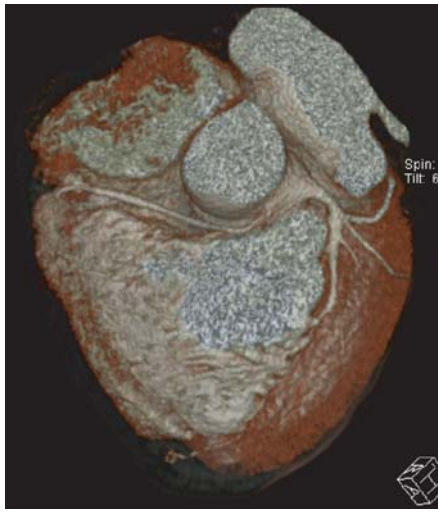
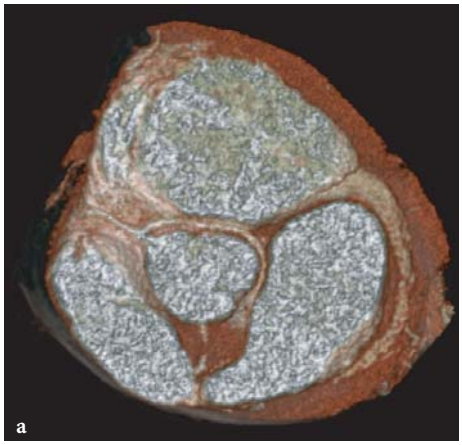


Fig. 7.56. 16-slice CT examination of an abnormal origin of the RCA from the left sinus of Valsalva, interarterial course. Note the narrowing of the proximal part of the RCA during its interarterial course. Volume-rendering reconstruction



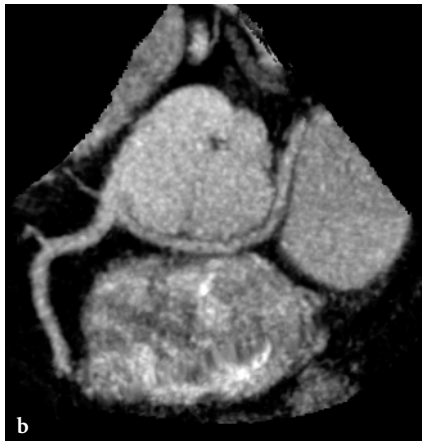


Fig. 7.55a,b. 16-slice CT examination of an abnormal origin of the CX from the right sinus of Valsalva, retro-aortic course. Displayed with volume rendering (**a**) and MIP reconstruction (**b**)

ations. Coronary artery fistulas should be treated surgically, with embolization or stenting of the communications. Even small fistulas with a hemodynamically low significant shunt should be closed to prevent further progression and complications. Spontaneous closure of a coronary artery fistula is extremely rare, but has been reported.

Some of the many types of possible fistulas are shown in Figure 7.58. As demonstrated in the case examples in Figures 7.59 and 7.60, 16-slice CT and 64-slice CT can reliably visualize a coronary fistula

7.8.4 Myocardial Bridges

Normally, the coronary arteries and their major branches course in the epicardial fat, but occasionally they course beneath the myocardium for various distances. Myocardial bridging is a congenital anomaly that is due to the failure of exteriorization of the primitive intratrabecular network of the affected coronary artery. Although this entity is almost always benign, cases of acute myocardial

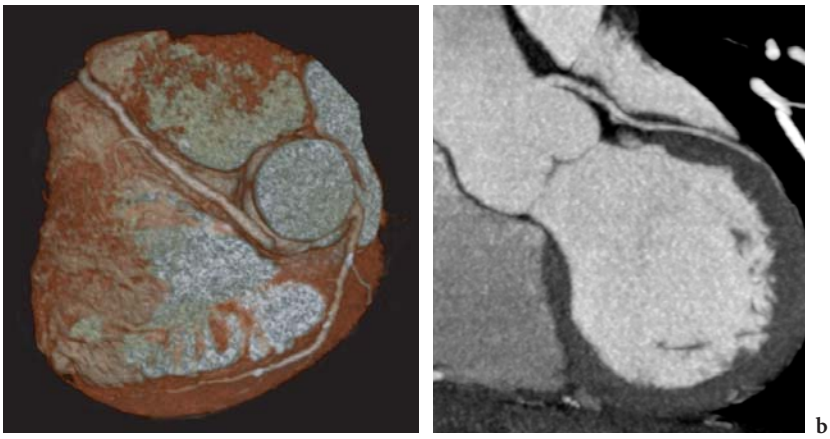


Fig. 7.57a,b. 16-slice CT examination of a clockwise malrotation of the aortic trunk, leading to an interarterial course of the RCA. Note the additional ectopic origin of the LMA above the left sinus of Valsalva. Visualization with volume-rendering reconstruction (**a**) and thin-MIP reconstruction (**b**) showing the aberrant origin of the LMA

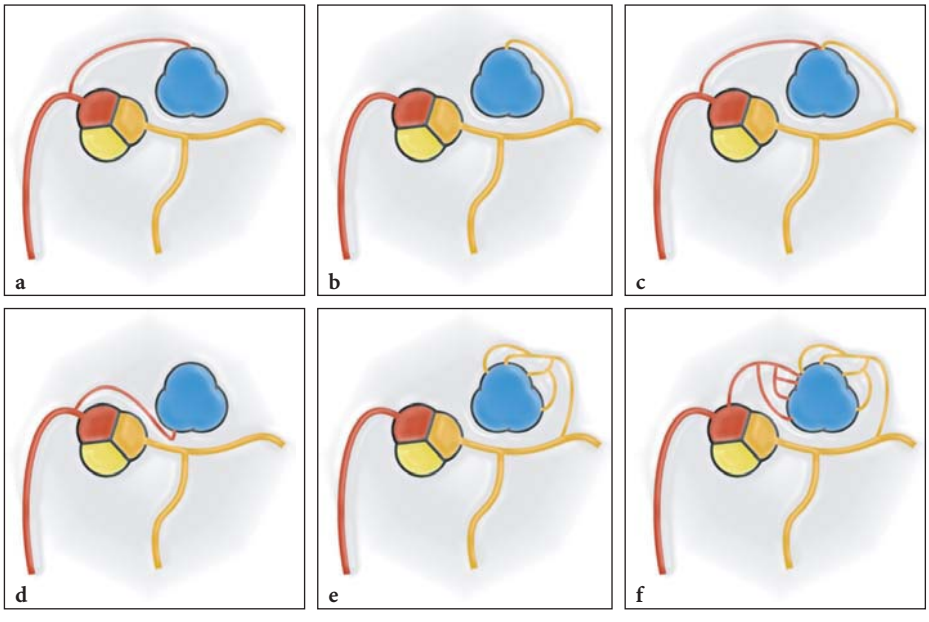
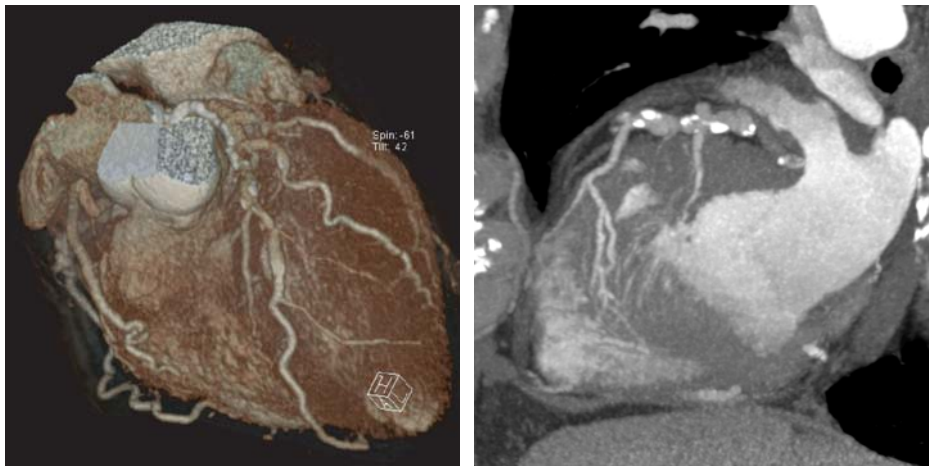


Fig. 7.58a–f. Possible coronary artery fistulas. **a** Fistula from the RCA to the pulmonary artery or other right-sided lumina. **b** Fistula from the LAD to the pulmonary artery or other right sided lumina. c Fistula from both the RCA and the LAD to the pulmonary artery or other right-sided lumina. **d** Fistula from the RCA to the pulmonary artery with an interarterial course. **e** Spider-like fistula from the LAD to the pulmonary artery or other right-sided lumina. **f** Spider-like fistula from both the LAD and the RCA to the pulmonary artery or other right-sided lumina



a b

Fig. 7.59a,b. 16-slice CT examination of a spider-like fistula from the LAD coursing through the interventricular septum and subepicardial to the right ventricle with multiple feeding vessels. Display with volume-rendering reconstruction viewed from the top (**a**) and with thin-MIP along a mid-ventricular short-axis section (**b**), showing the multiple fistulas through the interventricular septum to the right ventricle

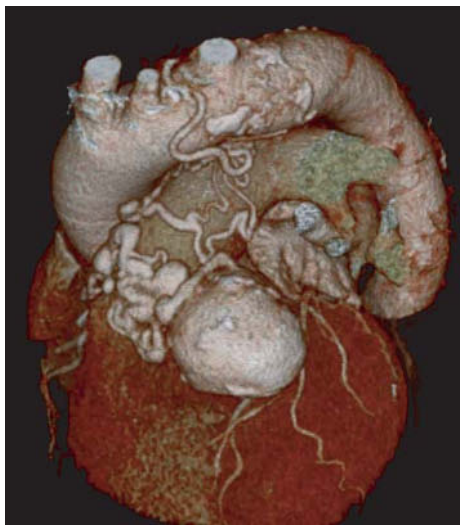


Fig. 7.60. Contrast-enhanced 16-slice CT coronary angiography with 16 × 0.75-mm collimation. Visualization of the left heart anatomy with 3D VRT reveals a complex and large fistula arising from the LAD and connecting to the ascending aorta. (Case courtesy of Hong Kong Sanatorium Hospital, China)

ischemia, cardiogenic shock, and even sudden death have been reported. Most patients do not have typical symptoms, such as angina, and their exercise stress tests are negative. Myocardial bridges are a common finding in autopsy. The incidence at postmortem examination is about 30–55%. In catheter angiography, they are detected in < 10% of patients. Any left coronary branch may be involved. The mid-segment of the LAD is by far the most common site. Males have a higher prevalence (70%). Myocardial bridges are also more common in patients with idiopathic left ventricular hypertrophy. Since the right ventricular systolic pressure is lower than the aortic pressure and the myocardial wall tension is smaller, myocardial bridges of the RCA system do not cause symptoms. The coronary artery is compressed during systole, and this can be demonstrated angiographically. Coronary blood flow occurs primarily during diastole. In myocardial bridges, blood flow may be hampered during increasing tachycardia. The presence of myocardial bridging distal to coronary lesions should be seriously considered to present a potential risk factor for intracoronary thrombus formation.

Unlike coronary angiography, multi-slice CT is capable of simultaneously depicting the course of the abnormal coronary vessel in direct relation to the myocardium. Reconstruction methods of choice

are thin perpendicular MPR or VRT. The systolic narrowing of the lumen can best be shown by reconstructing the data set at a systolic phase of the cardiac cycle and by comparing systolic and diastolic data sets. Multi-slice CT is thus a promising tool in detecting and depicting myocardial bridges, as demonstrated in Figure 7.61.

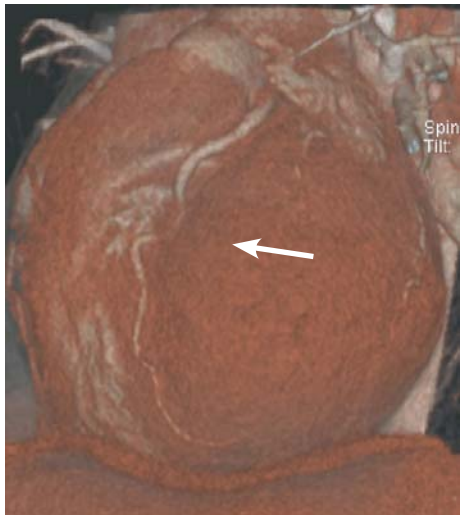


Fig. 7.61. 16-slice CT examination of a myocardial bridge (*arrow*) in the mid-segment of the LAD

7.8.5 Coronary Aneurysms

Coronary artery aneurysms are rarely characterized by abnormal dilatation of a focal portion or diffuse segments of the coronary artery. They are diagnosed incidentally at autopsy or at angiography in patients with symptoms of myocardial ischemia. The most frequent cause of aneurysms of the coronary arteries is atherosclerosis. In this regard, the dilatative form of atherosclerosis must be mentioned. An inflammatory pathogenesis that affects vessel wall modifications is Kawasaki syndrome, which is a clinically unspecific, acute, self-limited vasculitis of early childhood characterized by fever, bilateral nonexudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, and cervical lymphadenopathy. Coronary artery aneurysms may develop in about 25% of untreated children. Aneurysms can also occur in Takayasu arteritis. Vascular lesions causing aneurysms may result from chronic Chlamydia pneumoniae infection. Matrix metalloproteinases (MMPs) may be involved in the pathogenesis of arterial aneurysms due to increased proteolysis of the extracellular matrix of the arterial wall. Coronary artery aneurysms may also rarely develop in patients with systemic lupus erythematosus, sustained herpes virus infection, repetitive activation of virus-related antigens, suppressed immune state, Marfan syndrome, or multiple peripheral aneurysms. A frequent complication of coronary aneurysms is rupture, which mostly has a lethal outcome.

Multi-slice CT is able to depict the aneurysmal lumen as well as thrombus formation within the aneurysm. While 16-slice CT is capable of visualizing the dilated course of the vessels (Fig. 7.62), 64-slice CT accurately reveals the complex atherosclerotic changes (Fig. 7.63).

7.8.6 Coronary Dissection

Dissection of coronary artery is a quite rare but very acute entity. The most frequent cause is iatrogenic, i.e., catheter-induced. It highly important whether the aortic root is involved in the dissection process or not. Involvement of the aortic root may ultimately necessitate a complete replacement of the aortic-arch, perhaps with single re-implantation of the supra-aortic branches. Spontaneous coronary artery dissection is an important cause of acute coronary syndromes. It has been described to be associated with bodily stress, such as during sporting activities and even sexual intercourse. Presentation depends on the extent of the dissection and the location of the vessels involved. It can also occur predominantly in women during or after pregnancy. The association of coronary dissection with positive testing for anti-cardiolipin antibody or anti-phospholipid antibody, as seen in rheumatoid diseases, has been noted. The dissection membrane is best depicted in source images or thin-slice MPR perpendicular to the vessel course.

Dissections of the coronary arteries are seldom diagnosed by multi-slice CT, except the clinically

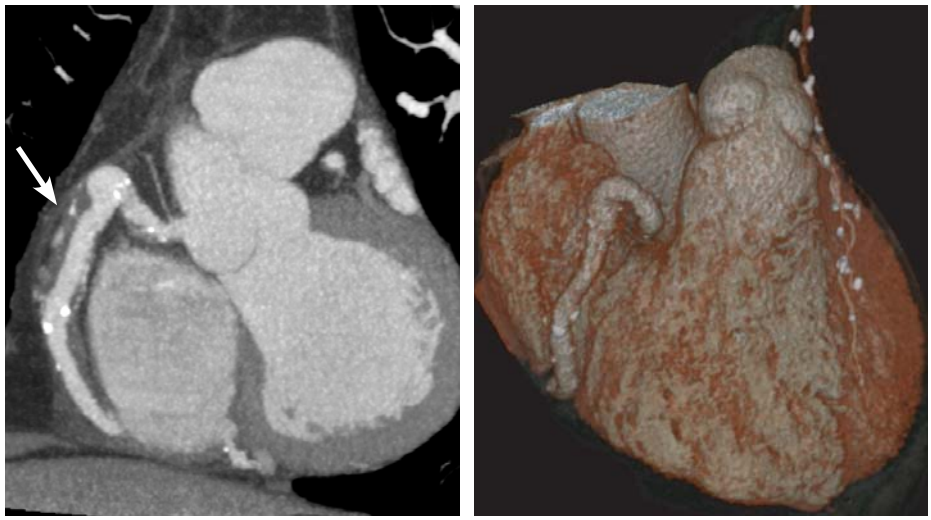


Fig. 7.62a,b. 16-slice CT examination of an aneurysm of the proximal RCA with extended thrombotic changes of the vessel wall. Displayed with thin-MIP projection (**a**) and volume-rendering reconstruction (**b**). The thrombus is visualized with thin-MIP (arrow in **a**). It is not shown in VRT display (**b**) but note a left IMA bypass to the distal LAD

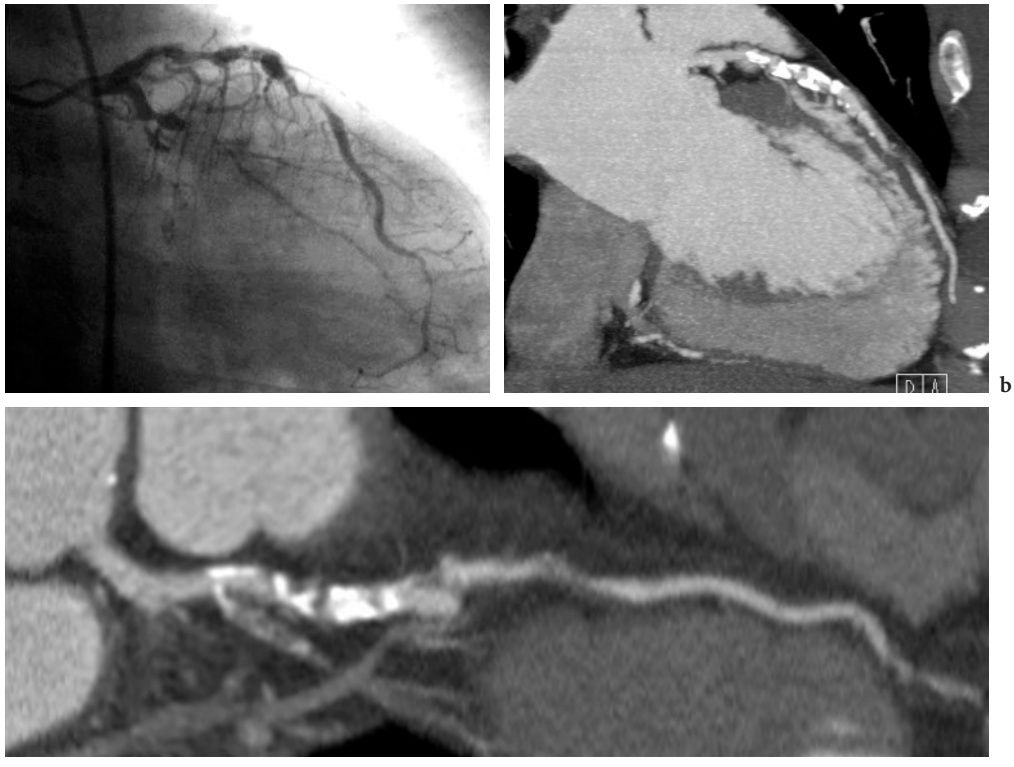


Fig. 7.63a–c. 64-slice CT examination of coronary aneurysms in the LAD. Conventional angiography demonstrates an abnormal course of the vessel (**a**) but does not allow visualization of the atherosclerotic changes. MPR along the long axis (**b**) and curved MPR (**c**) reveal the complex atherosclerotic lesions. (Case courtesy of Healthscan, Kuala Lumpur, Malaysia)

silent, chronic forms, which may be seen accidentally during CT scans for other medical conditions. The acute and clinically apparent forms are the domain of catheter angiography because of the option of immediate interventional therapy.

7.8.7 Coronary Vasculitis

Vasculitis-related lesions were mentioned previously. Only secondary alterations, such as aneurysms, stenoses, and acute and sub-acute closure of coronary arteries, can be depicted with multi-slice CT. As noted above, Kawasaki syndrome can cause coronary artery anomalies, including coronary fistulas and aneurysms in 20–25% of patients. In spite of early therapy, which consists of intravenous gamma-globulin and aspirin, coronary involvement may develop during the first years after diagnosis, even if there is early regress of the main underlying disease. In Kawasaki syndrome in particular, there may be peripheral occlusion of the coronary arteries and aneurysm. The specifically alteration of the arterial wall cannot be detected in multi-slice CT yet, due to the limited spatial resolution. There is no specific sign of coronary vasculitis in CT imaging. In other types of investigations, the value of multi-slice CT in detecting vasculitis has to be evaluated, e.g., in inflammatory thickening of the vessel wall.

7.8.8 Conclusion

In the last few years, multi-slice CT has become an alternative to catheter angiography. CTA is now the method of choice for detecting coronary arteries anomalies, fistulas, and aneurysms due to the 3D capability of this technique. Moreover, it is non-invasive, reproducible, and operator-independent. Especially in complex anomalies, if catheter angiography is not possible, multi-slice CT can accurately depict the anatomy of the heart and vessels. In contrast to catheter angiography, the thrombotic portion of aneurysms can be visualized with multi-slice CT. The new generation of CT scanners, with up to 64 slices, may improve image quality and resolution due to the smaller slice thickness and shorter breath-hold time. However, the ability of multi-slice CT to detect dissection and vasculitis of coronary arteries remains to be proven in future studies.

References

Chaitman BR, Lesperance J, Saltiel J, Bourassa MG (1976). Clinical, angiographic, and hemodynamic findings in patients with anomalous origin of the coronary arteries. Circulation 53: 122–131

Cheitlin MD, DeCastro CM, McAllister HA (1974). Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva. Circulation 50: 780–787

Click RL, Holmes DR Jr, Vlietstra RE, Kosinski AS, Kronmal RA (1989). Anomalous coronary arteries: Location, degree of atherosclerosis and effect on survival – A report from the Coronary Artery Surgery Group. J Am Coll Cardiol 13: 531–537

Dirksen MS, Langerak SE, de Roos A, Vliegen HW, Jukema JW, Bax JJ, Wielopolski PA, van der Wall EE, Lamb HJ (2002). Malignant right coronary artery anomaly detected by magnetic resonance angiography. Circulation 106: 1881–1882

Ishikawa T, Brandt PW (1985). Anomalous origin of the left main coronary artery from the right anterior aortic sinus: Angiographic definition of anomalous course. Am J Cardiol 55: 770–776

Kragel AH, Roberts WC (1988). Anomalous origin of either the right or left main coronary artery from the aorta with subsequent coursing between aorta and pulmonary trunk: Analysis of 32 necropsy cases. Am J Cardiol 62: 771–777

McConnell MV, Stuber M, Manning WJ (2000). Clinical role of coronary magnetic resonance angiography in the diagnosis of anomalous coronary arteries. J Cardiovasc Magn Reson 2: 217–224

Post JC, van Rossum AC, Bronzwaer JG et al (1995). Magnetic resonance angiography of anomalous coronary arteries. A new gold standard for delineating the proximal course? Circulation 92: 3163–3171

Reddy GP, Chernoff DM, Adams JR, Higgins CB (1998). Coronary artery stenoses: assessment with contrast-enhanced electron-beam CT and axial reconstructions. Radiology 208: 167–172

Roberts WC (1986). Major anomalies of coronary arterial origin seen in adulthood. Am Heart J 111: 941–963

Roberts WC, Siegel RJ, Zipes DM (1982). Origin of the right coronary artery from the left sinus of Valsalva and its functional consequences: Analyses of 10 necropsy patients. Am J Cardiol 49: 863–868

Ropers D, Moshage W, Daniel WG, Jessl J, Gottwik M, Achenbach S (2001). Visualization of coronary artery anomalies and their anatomic course by contrast-enhanced electron

beam tomography and three-dimensional reconstruction. Am J Cardiol 87: 193–197

Samarendra P, Kumari S, Hafeez M, Vasavada BC, Sacchi TJ (2001). Anomalous circumflex coronary artery: benign or predisposed to selective atherosclerosis. Angiology 52:521–526

Serota H, Barth CW III, Seuc CA, Vandormael F, Kern MJ (1990). Rapid identification of the course of anomalous coronary arteries in adults: The "dot and eye" method. Am J Cardiol 65: 891–898

Smith GT (1962). The anatomy of the coronary circulation. Am J Cardiol 9: 327–342

Taylor AM, Thorne SA, Rubens MP, Jhooti P, Keegan J, Gatehouse PD, Wiesmann F, Grothues F, Somerville J, Pennell DJ (2000). Coronary artery imaging in grown up congenital heart disease: Complementary role of magnetic resonance and x-ray coronary angiography. Circulation 101: 1670–1678

Van Ooijen PMA, Dorgelo J, Tukker W, Zijlstra F, Oudkerk M (2004). Visualization of anomalous coronary anatomy using multi-detector computed tomography (Abstract). Eur Radiol 14 (Suppl 2): 326

Yamanaka O, Hobbs RE (1990). Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn 21: 28–40

Yoshimura N, Hamada S, Takamiya M, Kuribayashi S, Kimura K (1998). Coronary artery anomalies with a shunt: evaluation with electron-beam CT. J Comput Assist Tomogr 22: 682–686