Coronary Artery Anomalies: Diagnosis and Classification based on Cardiac CT and MRI (CMR) – from ALCAPA to Anomalies of Termination

Koronararterienanomalien: Diagnostik und Klassifikation auf Basis der CT und MRT des Herzens – von ALCAPA bis Terminationsanomalie

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ABSTRACT

Background Coronary artery anomalies encompass a clinically and anatomically variable spectrum including physiological variants and pathophysiologically relevant anomalies. The majority of the variants has no hemodynamic relevance and is often detected accidentally. The recognition of the rare and relevant anomalies that cause either relevant shunt volumes leading to myocardial ischemia or ventricular tachyarrhythmias with the risk of sudden cardiac death is of major importance.

Methods This review is based on a literature search in PubMed conducted using the key words "coronary artery" and/or "anomaly" and/or "anomalous origin" and/or "myocardial bridging" and/or "coronary artery fistula" and/or "Bland-White-Garland" and/or "ALCAPA".

Results and Conclusion Coronary artery anomalies can be anatomically subdivided into anomalies of origin, course and termination. The method of choice for anatomical imaging is ECG-triggered or gated multislice CT (MSCT) that provides high spatial resolution and the capability of multiplanar reconstructions. It facilitates the delineation of the precise course of all three coronary arteries and thus allows for correct classification in the anatomical classification system of coronary artery anomalies. The strengths of cardiac magnetic resonance imaging (CMR) are the evaluation of cardiac morphology, myocardial tissue properties and myocardial function. Basic methods are the analysis of myocardial contraction and perfusion with and without pharmacologic stress. Furthermore, potential shunt volumes could be quantified by phase contrast imaging or volumetry.

Key points
- Coronary artery anomalies are subdivided into anomalies of origin, course and termination.
- The main imaging task is the differentiation of hemodynamically relevant anomalies from anatomic variants.
- The method of choice for anatomical imaging is MSCT, whereas structural and functional information is obtained by CMR

ZUSAMMENFASSUNG

Hintergrund Die Koronararterienanomalien umfassen ein klinisch variables und anatomisch variantenreiches Spektrum von Koronargefäßanlagevarianten bis hin zu pathophysiologisch relevanten Anomalien. Ein Großteil der Anlagevarianten ist hämodynamisch ohne Relevanz und wird häufig inzidentell nachgewiesen. Die große Bedeutung der Diagnostik von Koronararterienanomalien leitet sich aus den wenigen Anomalien ab, die pathophysiologisch entweder einen Shunt mit der Folge von Myokardischämien begründen oder die insbesondere ventrikuläre Tachyarrhythmien mit dem erhöhten Risiko eines plötzlichen Herztodes verursachen können.

Methode Die Übersichtsarbeit basiert auf einer Literaturrecherche in PubMed mit den Stichwörtern „coronary artery" und/oder „anomaly" und/oder „anomalous origin" und/oder „myocardial bridging" und/oder „coronary artery fistula" und/oder „Bland-White-Garland" und/oder „ALCAPA".

Ergebnisse und Schlussfolgerung Anatomisch kann man die Koronararterienanomalien in Abgangsanomalien, in Verlaufsanomalien und Terminationsanomalien unterteilen. Bildgebende anatomische Diagnostik der Wahl ist die EKG-getriggerte oder -gegatete Multislice-CT (MSCT) des Herzens, die alle drei genannten Koronargefäßabschnitte aufgrund einer hohen Ortsauflösung und der Möglichkeit der multiplanaren Reformatierungen darstellen kann und somit die Zuordnung zu vorgenannten Klassifikationskategorien ermöglicht.

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Die Bedeutung der Magnetresonanztomografie (MRT) liegt neben der strukturellen insbesondere in der funktionellen Beurteilung des Herzens. Grundlage hierfür ist die Analyse der myokardialen Kontraktilität und der Perfusion in Ruhe sowie unter medikamentöser Belastung. Ferner können potenzielle Shuntvolumina phasenkontrastbasiert oder planimetrisch quantifiziert werden.

Introduction

Coronary artery anomalies encompass a clinically and anatomically varied and complex spectrum of manifestations within the group of congenital heart defects [1 – 6]. Their clinical significance is derived from the possibility of myocardial ischemia and ventricular tachyarrhythmia (VT) with the potential complication of sudden cardiac death. In asymptomatic, young, athletic individuals, up to 15 % of cases of sudden cardiac death associated with increased physical activity were the result of an anomalous coronary artery origin [1, 2, 5 – 7]. The focus of this overview is the radiological, primarily CT-based diagnosis and systematic classification (▶ Table 1) of anatomical physiological variants and the resulting pathophysiologies.

The challenge when defining a classification system is to provide both intuitive criteria that can be implemented in the clinical routine and comprehensive representation of the spectrum of manifestations. The literature describes anomalies of origin, course and termination.

Under pathophysiological and clinical aspects, these categories are further subdivided into hemodynamically relevant anomalies, i. e., associated with shunts, ischemia, VT, and sudden cardiac death, and non-hemodynamically relevant or clinically asymptomatic coronary artery anomalies (▶ Table 1) [4, 6].

Anomalies of origin of the coronary arteries

Abnormal origin of the left coronary artery (LCA) from a pulmonary artery ALCAPA (anomalous origin of the left coronary artery from the pulmonary artery)/Bland-White-Garland syndrome

ALCAPA syndrome is a rare congenital coronary artery anomaly affecting 1/300 000 live births and represents 0.25 – 0.5 % of congenital heart defects [1, 2, 8, 9]. It is characterized by an anomalous origin of the left coronary artery (LCA) from the pulmonary trunk (PT) or from a pulmonary artery (PA) (▶ Table 1, ▶ Fig. 1) [8 – 11]. This origin variant initially has no effect on the hemodynamics of the coronary arteries in the prenatal and early neonatal phase. The blood pressure conditions between the systemic circulation and the pulmonary arterial circulation are equalized by the patent ductus arteriosus. This initially ensures a forward flow in the LCA from the PA (▶ Fig. 2a). Physiological occlusion of the ductus arteriosus lowers the pressure in the pulmonary arterial circulation. This causes a decreasing flow rate in the LCA that ultimately results in a reverse of the flow of oxygenated blood in the direction of the PA. A left-right shunt is created in that oxygenated blood

▶ **Table 1** Overview of the coronary artery anomalies [4, 6].

coronary artery anomalies	
hemodynamically relevant	**non-hemodynamically relevant**
anomalies of origin	
anomalous origin of the LCA from the PA (ALCAPA/Bland-White-Garland syndrome)	high takeoff (> 1 cm above the sinotubular junction)
anomalous origin of a CA from the contralateral CS (ACAOS) with interarterial course	origin outside the CS ▪ posterior noncoronary sinus ▪ ascending aorta (e. g. high takeoff) ▪ left, right ventricle ▪ aortic arch, descending aorta, bronchial artery ▪ subclavian artery, internal thoracic artery, carotid artery
	multiple ostia (e. g. lack of LCA main branch)
	ACAOS with ▪ Retroaortic ▪ Transseptal/subpulmonic ▪ Prepulmonic course
anomalies of course	
myocardial bridge	
	duplication
	crossing of coronary artery segments
anomalies of termination	
coronary artery fistula (CAF)	coronary arcade
	extracardiac systemic termination

ACAOS: anomalous origin of a coronary artery from the opposite sinus; ALCAPA: anomalous origin of the left coronary artery from the pulmonary artery; CA: coronary artery; CS: coronary sinus; LCA: left coronary artery; PA: pulmonary artery.

flows through the LCA to the PA (▶ **Fig. 2b, c**) [8, 9]. There are two types of ALCAPA syndrome: infantile (▶ **Fig. 2b**) and adult (▶ **Fig. 2c**) [8, 9].

The adult type is characterized by the compensatory formation of collaterals between the RCA that physiologically originates from the aorta and the LCA [8, 9, 12]. These collaterals can compensate for the shunt volume to varying degrees in individual cases (▶ **Fig. 2**). The spectrum of clinical manifestations ranges from an asymptomatic course to decompensation of the coronary arterial collateral circulation with the result of clinical revealing of a potentially underlying, previously chronically subclinical ischemia. Ischemia can result in ischemic cardiomyopathy, secondary
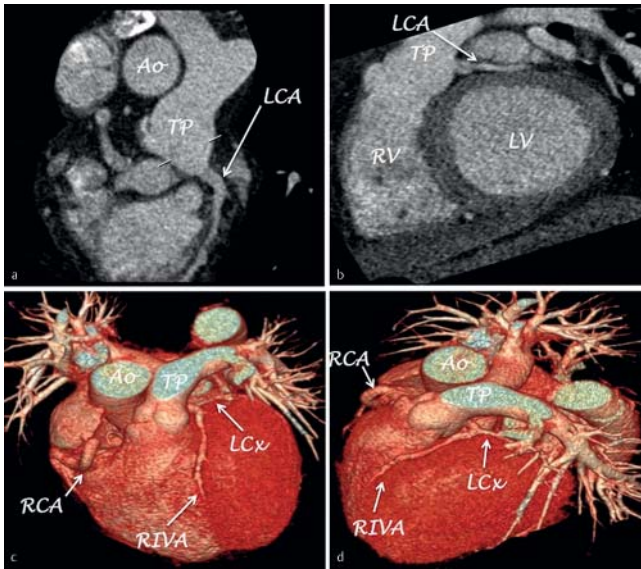
► **Fig. 1** ALCAPA/Bland-White-Garland syndrome; **a, b** MPR of a CT coronary angiography with anomalous origin of LCA from the TP; **c, d** VRT with anomalous origin of LCA from the TP. (ALCAPA: anomalous origin of the left coronary artery from the pulmonary artery; Ao: aorta; LCA: left coronary artery; LCx: left circumflex artery; MPR: multiplanar reconstruction; RCA: right coronary artery; TP: truncus pulmonalis; VRT: volume-rendering technique).

mitral valve insufficiency, or ventricular arrhythmia with the latter increasing the risk of sudden cardiac death [1, 8, 11, 13].

In the infantile type collaterals do not form between the RCA and LCA (► **Fig. 2**). Symptoms in the form of paleness, pronounced sweating, dyspnea, and chest pain begins in the 4th to 8th week of life [8, 9]. The lack of oxygenated blood supply to the LV myocardium results in myocardial infarcts (► **Fig. 2d-g**). These can cause secondary mitral valve insufficiency and ischemic cardiomyopathy with cardiac insufficiency. Without corrective surgery, the mortality rate in the first weeks to months of life is 90 % [8, 11, 12].

The imaging modalities of choice are multislice CT (MSCT) and magnetic resonance imaging (MRI) of the heart (► **Fig. 1, 2**). The primary visible morphological characteristics of ALCAPA syndrome are detection of an anomalous origin (► **Fig. 1**) and the left-right shunt with retrograde flow from the LCA into the PA. MSCT with its high spatial resolution and the possibility of multiplanar reconstruction allows direct morphological visualization of the anatomical variations in origin (► **Fig. 1**) [8, 14]. Proximal coronary vessel segments and variations in origin from the thoracic aorta or the pulmonary trunk can be visualized with MR angiography. However, diagnosis via MSCT is more robust with respect to artifacts. Functional evaluation is performed via MRI with cine sequences that can detect the reverse of flow in the LCA comparably to catheter coronary angiography [8, 15]. Secondary visible morphological characteristics of the adult type are a dilated, elongated RCA (► **Fig. 2**) and dilated intercoronary collateral arteries that can be better visualized with MSCT than MRI. Moreover, LV dilatation, regional LV wall motion abnormalities, and secondary mitral valve insufficiency should be mentioned in this connec-

tion and are best detected using MRI. Motion abnormalities are detected by cine sequences and underlying infarct areas by late gadolinium enhancement (LGE) sequences (► **Fig. 2**) [8, 15, 16].
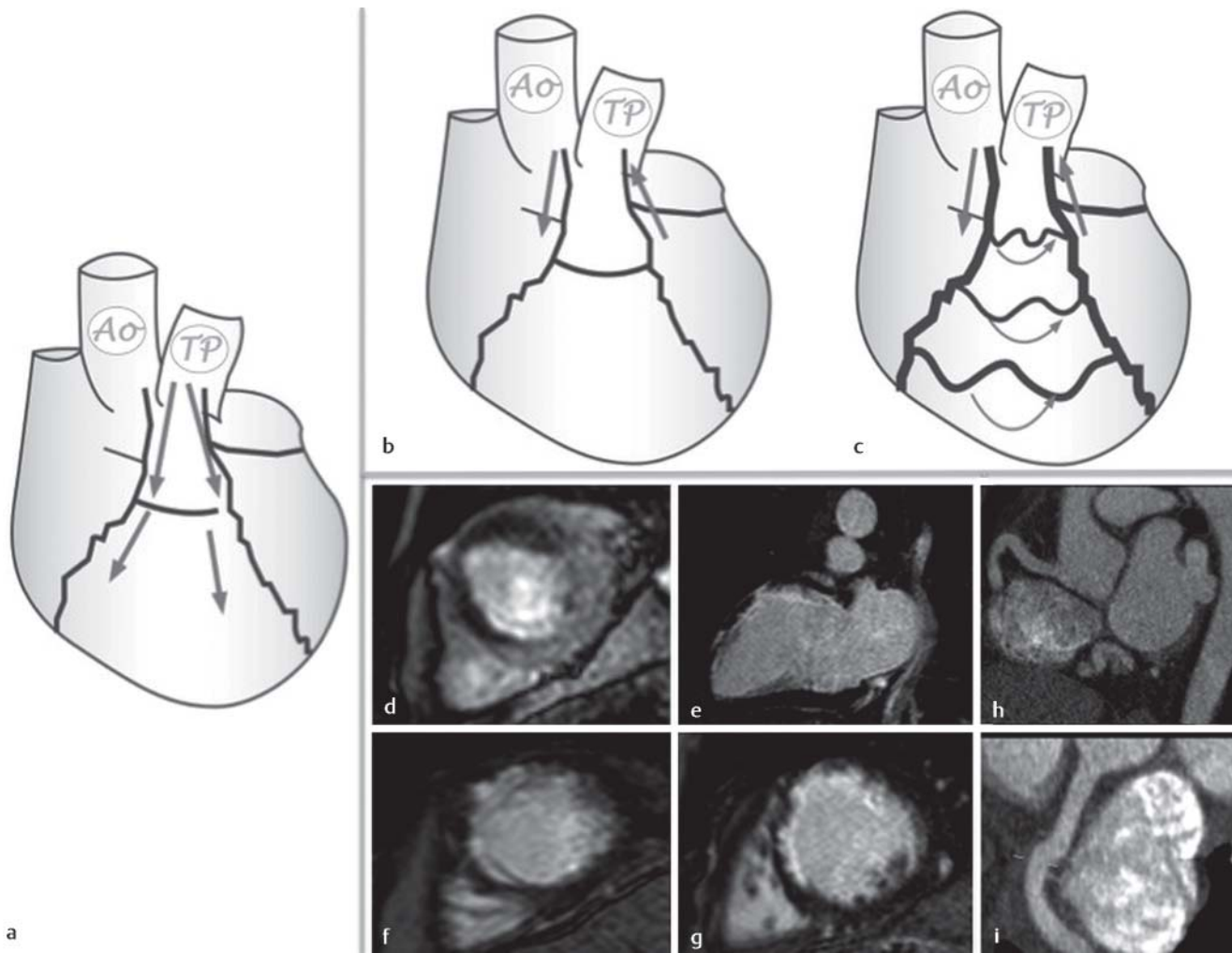
Corrective surgery is indicated in the case of primary diagnosis in newborns and in the case of the adult type in combination with the detection of limited LV function or extensive subendocardial LGE (► **Fig. 2**) [2, 8, 17, 18]. For surgical treatment, corrective techniques based on 2-coronary-artery systems are preferred. These include the reimplantation of the LCA in the aorta ("coronary button transfer"), which is favored in newborns, the related Takeuchi maneuver with a baffle between the pulmonary arterial ostium and aortic anastomosis in the pulmonary arterial vascular lumen, and aortic coronary bypass operation in combination with ligature of the anomalous LCA origin from the PA [1, 8, 17 – 19].

Abnormal origin of a coronary artery from the contralateral or noncoronary sinus ACAOS (anomalous origin of a coronary artery from the opposite sinus) and associated variations in vessel course: interarterial, retroaortic, transseptal/subpulmonic, prepulmonic

There are four different types of anomalous origin of a coronary artery from the contralateral coronary sinus (ACAOS): Origin of the LCA from the right coronary sinus (RCS) (► **Fig. 3**), origin of the right coronary artery (RCA) from the left coronary sinus (LCS) (► **Fig. 3**), anomalous origin of the left circumflex artery (LCx) or the ramus interventricularis anterior (RIVA) from the RCS (► **Fig. 4**) and origin either of the RCA or the LCA from the noncoronary sinus as the rarest variant (► **Fig. 3**). In this connection, the particular coronary artery can arise at the opposite coronary sinus from a separate ostium (► **Fig. 3**) or with the original coronary vessel from a shared coronary artery trunk (► **Fig. 3**) [3, 4, 20].

The incidence of ACAOS of the two large coronary arteries is 1.07 % with a total incidence of all coronary artery anomalies of approx. 5.64 % [1, 2, 5, 6]. The incidence is 0.92 % for an anomalous origin of the RCA from the LCS and 0.15 % for an origin of the LCA from the RCS [5, 6]. With a rate of 0.67 %, the LCx is the vessel that arises most frequently from the RCS (► **Fig. 4**) [4 – 6].

In association with ACAOS, the course of the coronary artery crosses to the contralateral half of the heart in order to supply its corresponding vessel territory [3, 4, 20]. In this connection, four different courses that can be applied to each of the four previously mentioned types of ACAOS are known (► **Fig. 4**). These include the interarterial course (► **Fig. 4 – 6**), the retroaortic course (► **Fig. 4, 7**), the transseptal or subpulmonic course (► **Fig. 4**), and the prepulmonic course (► **Fig. 4**) (► **Table 1**) [4]. A pathophysiological specification is added to this anatomical classification in that the course variants are classified as hemodynamically relevant and non-hemodynamically relevant types [4]. The non-hemodynamically relevant course types include the retroaortic (► **Fig. 4, 7**), transseptal (► **Fig. 4**), and prepulmonic course (► **Fig. 4**) (► **Table 1**) [4]. In contrast, the interarterial course can be associated with sudden cardiac death and is classified as hemodynamically relevant (► **Table 1**, ► **Fig. 4 – 6**) [1, 2, 4, 20, 21].

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► **Fig. 2** Pathophysiology of ALCAPA syndrome of infantile and adult type; **a** prenatal and early postnatal coronary arterial circulation: patent ductus arteriosus; Psyst = Ppul; antegrade blood flow in LCA and RCA; **b** infantile type: occlusion of ductus arteriosus; Psys > Ppul: conversion of blood flow in LCA towards TP; absent collaterals between RCA and LCA; **c** adult type: Psys > Ppul; conversion of blood flow in LCA; collaterals between RCA and LCA with dilatation of RCA; **d** MR perfusion: delay of perfusion of anteroseptal wall; **e–g** IR-LGE sequences with subendocardial infarction anteroseptal; **e** 2Ch; **f** SA apical; **g** SA midventricular; **h**, **i** CT-MPR of dilated RCA in adult type; (DAB: ductus arteriosus Botalli; Ppul: pulmonary arterial pressure; Psys: systemic arterial pressure; IR-LGE: inversion-recovery late gadolinium enhancement, further abbreviations Fig. 1).

An intramural course of the particular coronary vessel that is variable with respect to passage length, i. e., a course embedded in the vascular wall of the ascending aorta, can additionally be detected on the basis of the critical hemodynamics (► **Fig. 3**). This course that is potentially combined with the interarterial type of course can be considered a pathophysiologically relevant cofactor for the resulting critical hemodynamics [20, 21]. Additional anatomical cofactors contributing to critical hemodynamics are a slit-like ostium and an acute angle takeoff from the coronary sinus (► **Fig. 6**) [20, 21].

The etiological cause of sudden cardiac death with a close association with increased physical activity is an anomalous origin of the coronary artery in up to 15 % of cases [1, 5 – 7]. An interarterial course is detected as the cause in 80 % of cases of sudden cardiac death and a simultaneously confirmed anomalous origin of the coronary artery in young athletes [1]. Additional clinical manifestations can be chest pain, arrhythmia, LV dysfunction, or

stress-induced syncope [1]. Patients who survive sudden cardiac death or who present with the specified symptoms should be diagnosed with respect to anomalous origin and course of the coronary artery [1].

The diagnostic modality of choice is either MSCT coronary angiography or cardiac MRI with a MRA of the coronary arteries with distal segments of the coronary arteries and an anomalous vessel course also being able to be detected with the former method [1, 2]. Information regarding the prevalence of coronary artery anomalies and in particular ACAOS is based on the results of coronary angiography series. Prior to the further development of MSCT, catheter coronary angiography was the diagnostic modality of choice for detecting coronary artery anomalies. However, numerous studies have shown that MSCT coronary angiography is superior to conventional angiography in the detection of coronary artery anomalies; only approx. 53 – 55 % of anomalies detected by MSCT coronary angiography could be detected by catheter angio-

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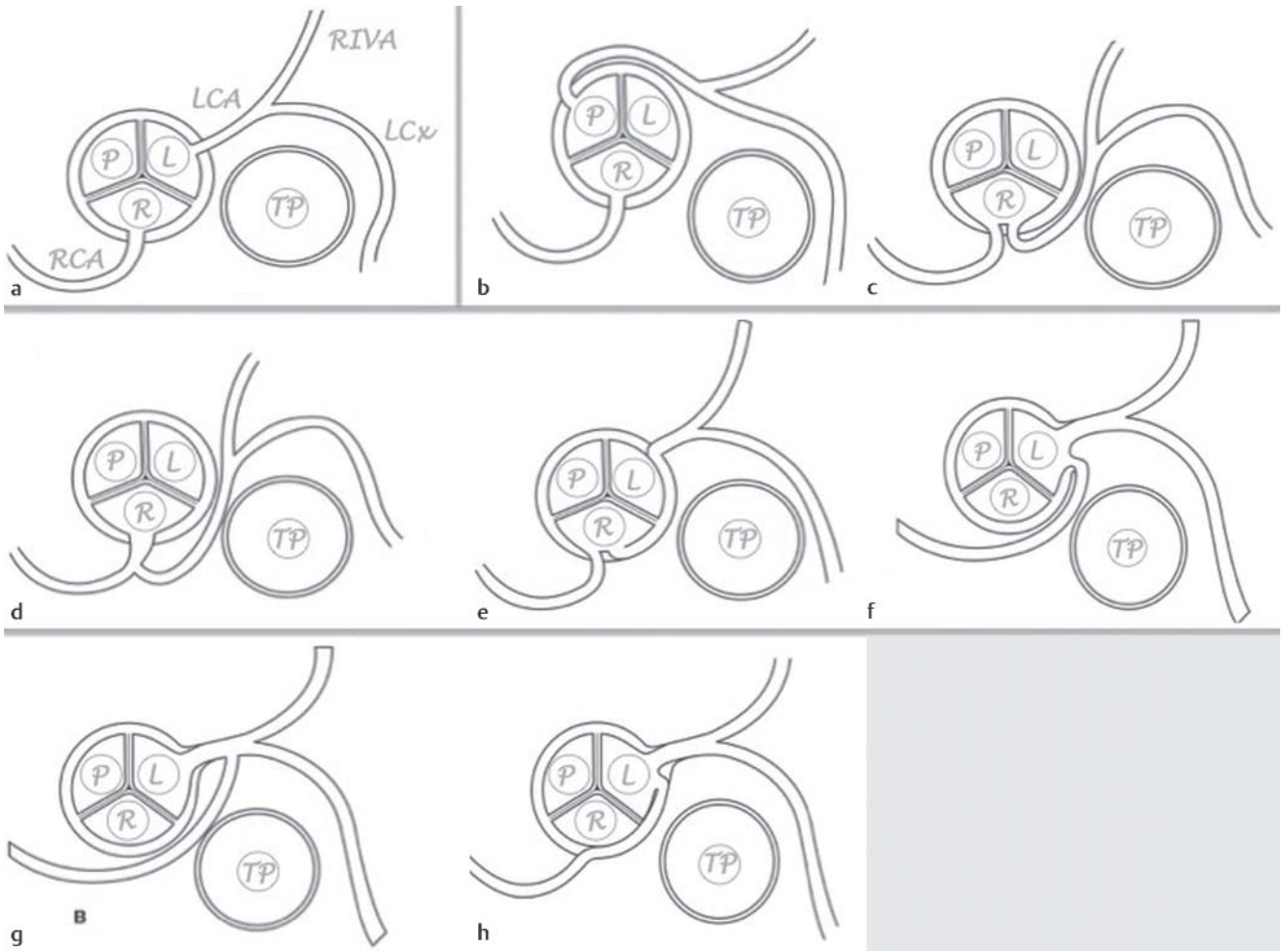
▶ **Fig. 3** Variants of coronary arterial anomalies of origin at coronary sinus; **a** normal anatomy as reference; **b** anomalous origin of LCA from posterior sinus; **c–h** ACAOS; **c–e** anomalous origin of LCA from RCS with interarterial course: c: two ostia with separate origin of LCA and RCA from RCS; **d** one ostium and common origin; **e** intramural course in the wall of the aorta; **f–h** anomalous origin of RCA from LCS with interarterial course; variants **f–h** comparable to **c–e**; (ACAOS: anomalous origin of a coronary artery from the opposite sinus; LCS: left coronary sinus; RCS: right coronary sinus; further abbreviations Fig. 1).

graphy [3, 22]. As in ALCAPA syndrome, MRI is most suitable for functional evaluation. Myocardial ischemia affecting surgical decision making can be detected with MR perfusion of the myocardium during pharmacologically induced stress MRI [1, 2]. To complete the overview of the available noninvasive imaging modalities for detecting myocardial ischemia, dobutamine-stress echocardiography and nuclear medicine methods, e. g. in the form of myocardial scintigraphy, should be mentioned in addition to stress MRI of the heart [23].

Surgical revascularization methods and percutaneous coronary interventions (PCI) are available for treating hemodynamically relevant origin and course anomalies [1, 20, 24, 25]. Surgical treatment options include coronary bypass surgery which is increasingly less preferred because of potential complications such as stenoses and occlusions of the bypass due to competitive flow from the anomalous coronary artery that is patent if ligature is not performed [24]. Additional surgical methods are coronary re-implantation of anomalous origins of the coronary artery, anterior or lateral pulmonary artery translocation, or surgical exposure of

an intramural coronary artery segment running in the aortic wall (unroofing) [1, 20].

According to the recommendations of the American College of Cardiology/American Heart Association from 2008, surgical therapy is indicated in the case of ACAOS with an interarterial course. In addition to the interarterial course in the case of an anomalous origin of the LCA from RCS (▶ **Fig. 5**), it is necessary to detect an associated ischemia in the corresponding vessel territory in the case of an anomalous origin of the RCA from the LCS (▶ **Fig. 6**) [1, 2].

Additional anomalies of origin without hemodynamic relevance

This includes the detection of multiple coronary ostia within the coronary sinus, resulting, for example, in a separate origin of the RIVA and the LCx from the LCS and the so-called "high takeoff", "high origin" (▶ **Table 1**) [3, 4]. The latter anomaly of origin is defined as the origin of the left or right coronary ostium > 1 cm above the sinotubular junction, more commonly observed in the

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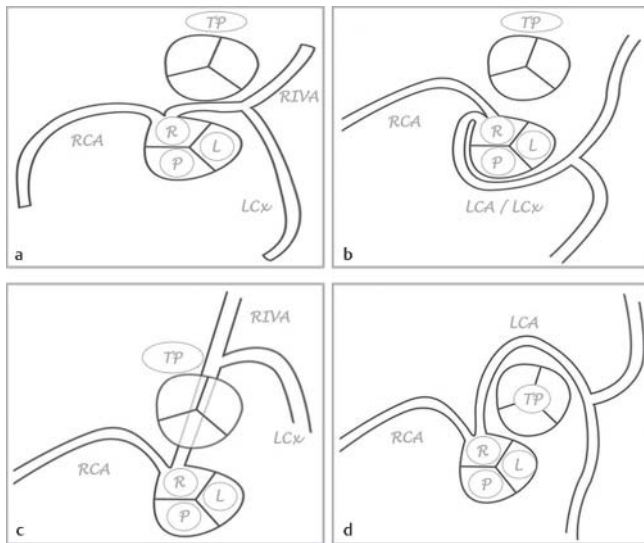
► **Fig. 4** Variants of course in ACAOS (LCA from RCS); **a** interarterial course; **b** retroaortic course; **c** transseptal/subpulmonic course; **d** prepulmonic course; (ACAOS: anomalous origin of a cor-

case of the RCA and associated with the potential detection of a bicuspid aortic valve [3]. This anomaly of origin has no hemodynamic relevance. However, its accidental detection plays an important role in CT coronary angiographic evaluation prior to planned replacement of the aortic valve or ascending aorta [3].

Anomalies of course of the coronary arteries

Anomalies of course include myocardial bridging (► **Table 1,** ► **Fig. 8**), the crossing of two coronary vessel segments from different coronary arteries, and the duplication of a coronary artery [4, 6]. The later anomaly of course is most commonly detected along the RIVA. It is characterized by a short LCA vascular branch ending in the sulcus interventricularis anterior (SIVA) just before reaching the apex and a longer accessory vascular branch that arises from the LCA or RCA and supplies the apex. This anomaly of course has no hemodynamic relevance but plays an important role as an accidental finding in the CT angiographic evaluation prior to planned aortocoronary bypass surgery [4]. As distinguished from the interarterial course, the non-hemodynamically relevant coronary arterial courses in the form of the retroaortic (► **Fig. 4, 7**), transseptal (► **Fig. 4**), and prepulmonic (► **Fig. 4**) courses were already specified as further anomalies of course in combination with ACAOS.

Myocardial bridging

The coronary arteries follow a subepicardial course. A myocardial bridge is defined by a coronary artery segment that is surrounded by heart muscle tissue in an intramyocardial, circular fashion (► **Table 1**, ► **Fig. 8**) and can be compressed during systolic contraction [4, 26 – 30]. The most common location is the middle segment of the RIVA [4, 26 – 30]. There is a clear discrepancy between the data regarding the prevalence of myocardial bridges

from catheter angiography series (0.5 – 2.5 %) and autopsies (15 – 85 %) [4, 26, 28].

In most cases a myocardial bridge is a benign physiological variant without clinical symptoms [26 – 30]. However, due to a reduced blood flow rate in the coronary vessel triggered by vessel compression during systolic myocardial contraction and by delayed vessel relaxation during diastole, myocardial ischemia, myocardial infarcts, ventricular arrhythmias and sudden cardiac death can occur [26, 30]. The extent of this coronary vessel compression is affected by anatomical concomitant factors such as length and location of the myocardial bridge, thickness and depth of the myocardial band forming the bridge and the simultaneous presence of myocardial hypertrophy. In studies regarding quantitative coronary angiography and intracoronary Doppler ultrasound, reference values for hemodynamically relevant myocardial bridges were created. In detail, these are reduced systolic and mid-diastolic average lumen diameters, elevated average systolic and diastolic peak flows and a reduced distal coronary flow reserve. The percentage of coronary vessel lumen reduction during systole is divided into 3 levels: Level 1 < 49 %, level 2 50 – 74 %, level 3 > 75 % lumen reduction [31, 32].

In addition to hemodynamic considerations, the detection of a myocardial bridge plays a role in the evaluation prior to planned aortocoronary bypass surgery since this complicates surgical access to the coronary segment embedded in the myocardium [30]. In the literature myocardial bridges are detected most frequently accidentally via catheter coronary angiography in which the characteristic "milking effect", i. e., varying width of the coronary lumen depending on the cardiac cycle, is observed or via MSCT of the heart [30]. An advantage of MSCT with the option of curved and multiplanar reconstruction is the simultaneous visualization of the vascular lumen, the vessel wall, and the relationship of the coronary vessel to the surrounding myocardium and to the cavities of the heart [4, 26, 28, 30].

Since a varying lumen width of a coronary segment during the cardiac cycle can also be detected with MSCT in that a reconstruction can be created during end-systole in addition to the established reconstruction window during end-diastole [4].

The treatment of choice in symptomatic myocardial bridges is the pharmacological application of beta blockers with a negative inotrope and chronotrope effect to reduce vascular lumen constriction. In addition to the pharmacological effects during systole, the early diastolic blood flow is reduced and the systolic/diastolic flow ratio is improved. In the case of contraindications for the application of beta blockers and vasospasms present at the same time as myocardial bridging, the application of calcium receptor antagonists is indicated [31].

Surgical therapy is usually not necessary for the typically benign physiological variant without clinical symptoms [26 – 30]. Surgical exposure of the intramyocardial vascular segment by myotomy or bypass surgery is reserved for the few clinically symptomatic and hemodynamically relevant or pharmacologically refractory myocardial bridges [30].
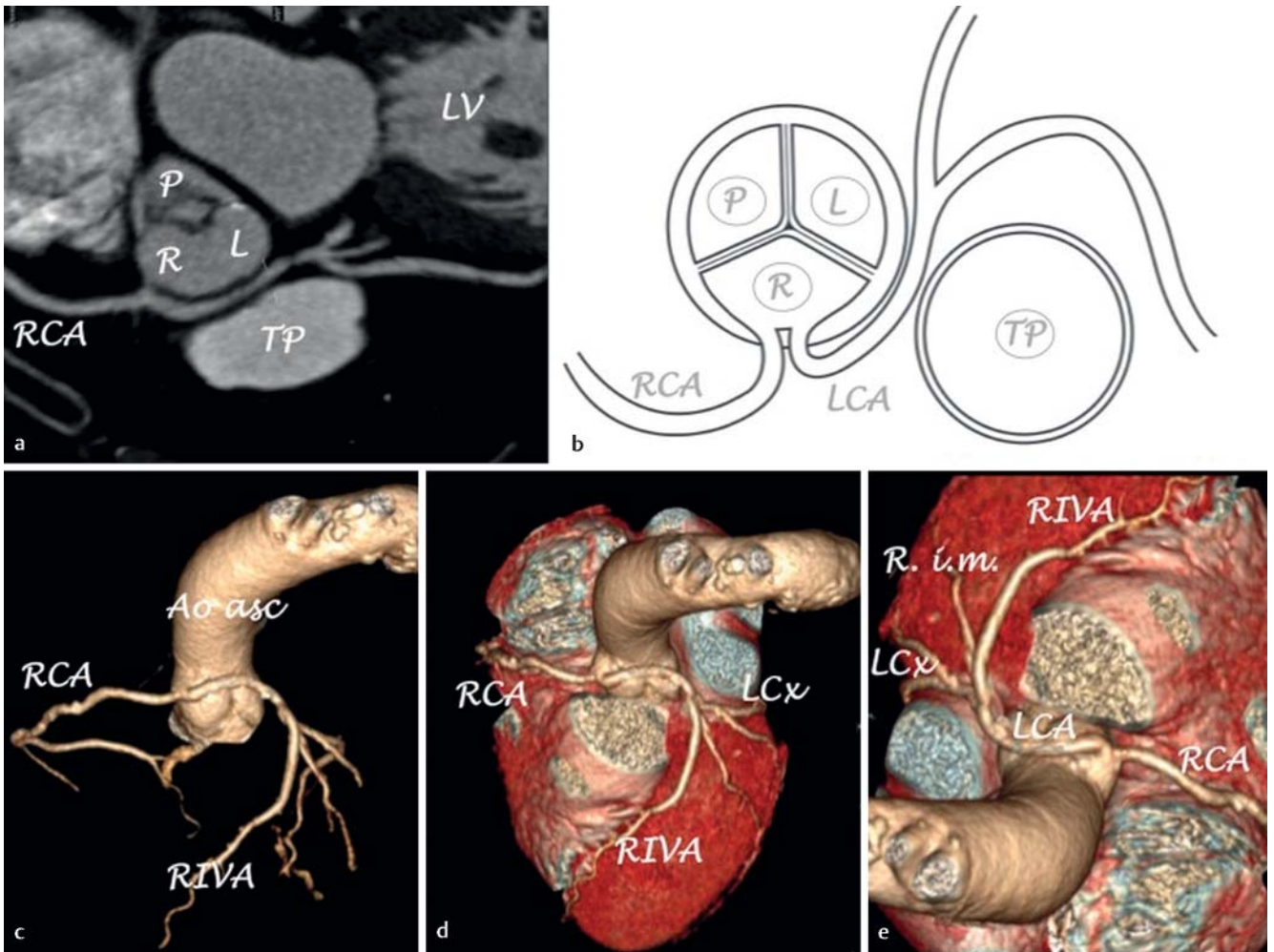
▶ **Fig. 5** Anomalous origin of LCA from RCS with interarterial course; **a** curved reconstruction; **b** scheme; **c** CT-VRT of coronary artery system; **d, e** CT-VRT of the heart in varying views; (Ao asc: ascending aorta; CT: computed tomography; L: left coronary sinus; LCA: left coronary artery; LCx: left circumflex artery; LV: left ventricle; P: posterior sinus; R, RCS: right coronary sinus; RCA: right coronary artery; R.i. m.: ramus intermedius; RIVA: Ramus interventricularis anterior; TP: Truncus pulmonalis; VRT: volume-rendering technique).

## Anomalies of termination of the coronary arteries

Anomalies of termination include hemodynamically relevant coronary arterial fistula (CAF) (▶ **Fig. 9**), which will be discussed in detail in the following, as well as the coronary arcade, which is defined as an angiographically detectable communicating vascular network between the LCA and RCA, and the extracardiac systemic termination of coronary arteries without a dilated and elongated vessel course, which is to be delimited from a CAF (▶ **Table 1**) [4, 6].

Coronary artery fistula (CAF)

CAF is defined by an abnormal termination of a coronary artery. Toward its physiological termination in the myocardial capillary vascular bed, there is direct communication with a cavity of the heart, with veins of the systemic circulation, the lungs or the heart, or with the pulmonary arteries [1, 3, 4, 33 – 40]. The coronary angiographic incidence of CAF is approx. 0.1 – 0.2 % [1, 4] and

represents approx. 0.13 % of congenital heart defects [33]. The RCA is the most commonly affected vessel (60 % versus LCA 40 %) and a CAF drains primarily into the RV (45 %) followed by communication with the RA (25 %) or a PA (15 %) [4, 33].

Pathophysiologically, the location of the vessel termination before the origin of the CAF is highly relevant. In the case of communication with a vascular structure of the right half of the heart, an extracardiac left-right shunt is present and in the case of drainage to the left half of the heart, a left-left shunt, comparable with aortic insufficiency, is formed [4, 33]. Communication with a cardiac cavity or vascular structures of the low pressure system results in dilation and elongation of the coronary vessels representing a diagnostic characteristic in imaging (▶ **Fig. 9**). This results in a steal phenomenon or a shunt with bypass of the capillary vascular bed to be physiologically supplied by the affected coronary artery with the potential consequence of myocardial ischemia [2 – 4]. Additional associated complications can be endarteritis, LV dysfunction, ventricular arrhythmias, coronary vessel dissection or rupture [1].

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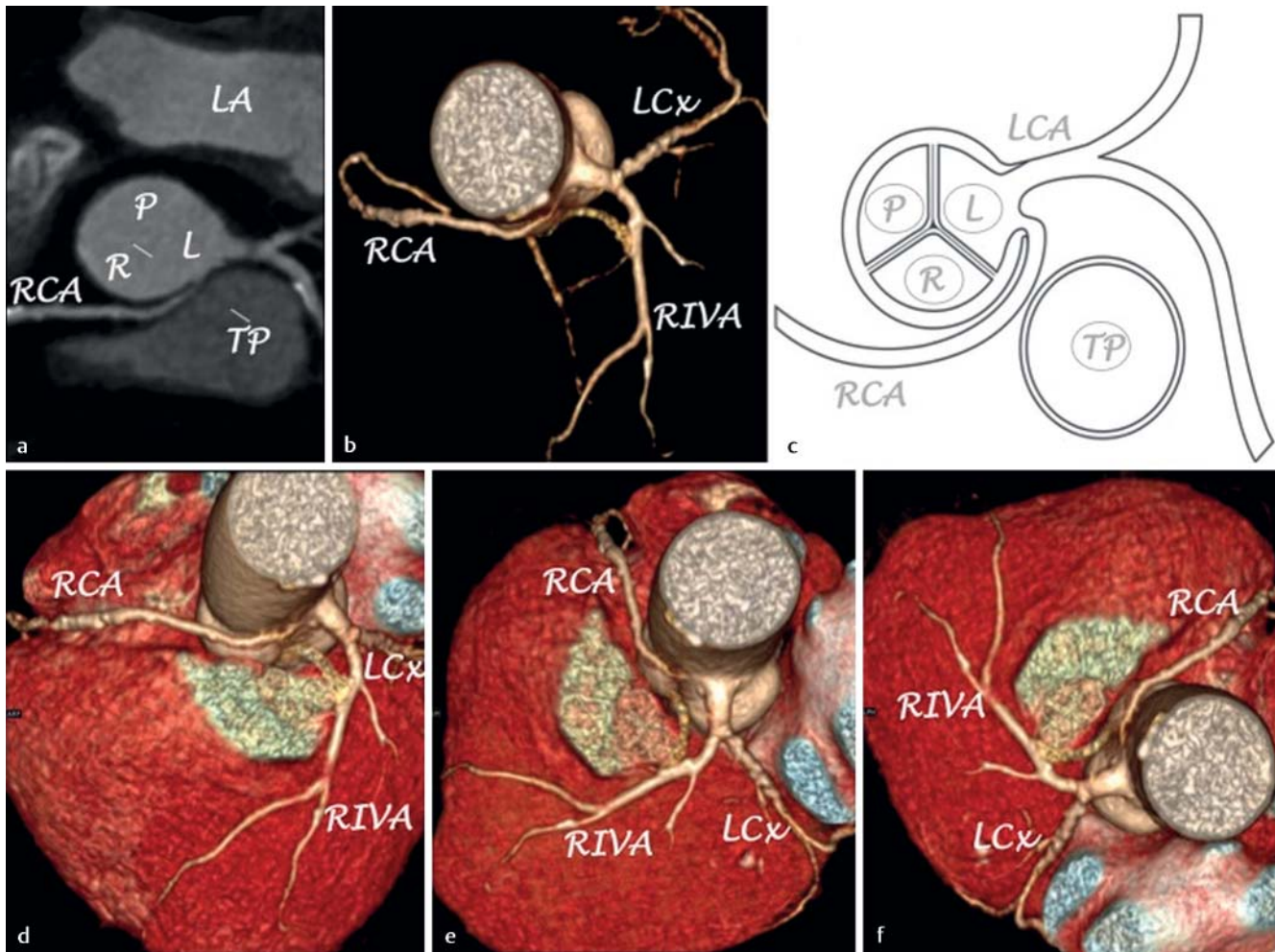
▶ **Fig. 6** Anomalous origin of RCA from LCS with interarterial course; **a** curved reconstruction; **b** CT-VRT of coronary artery system; c scheme; **d–f** CT-VRT in varying views; (LCS: left coronary sinus; further abbreviations Fig. 5).

CAF can be detected with echocardiography, MSCT, MRI, and catheter angiography [1, 2]. With high spatial resolution, MSCT is suitable for the detection of small CAFs and simultaneous three-dimensional visualization of the potential communicating anatomical structures listed above via multiplanar reconstruction. Echocardiography-based follow-up every 3 to 5 years is recommended in the case of small, clinically asymptomatic CAFs [1, 2].

The indication for treatment via surgical correction or catheter embolization methods is determined at specialized centers in the case of large CAFs or in the case of small and medium CAFs in combination with the indicated symptoms or complications [1, 2].

In summary, coronary artery anomalies can be anatomically categorized as anomalies of origin, course, and termination and include a clinically variable and anatomically varied spectrum that ranges from physiological variants to pathophysiologically and hemodynamically relevant anomalies.

The major importance of diagnostic imaging is the sensitive detection of rarer, hemodynamically relevant coronary artery anomalies including the ALCAPA/Bland-White-Garland syndrome, the interarterial coronary arterial course in ACAOS, coronary arterial fistulas, and rarely also myocardial bridges.

The imaging modality of choice for anatomical diagnosis is MSCT of the heart, while MRI is important for the structural as well as in particular the functional evaluation of the heart.
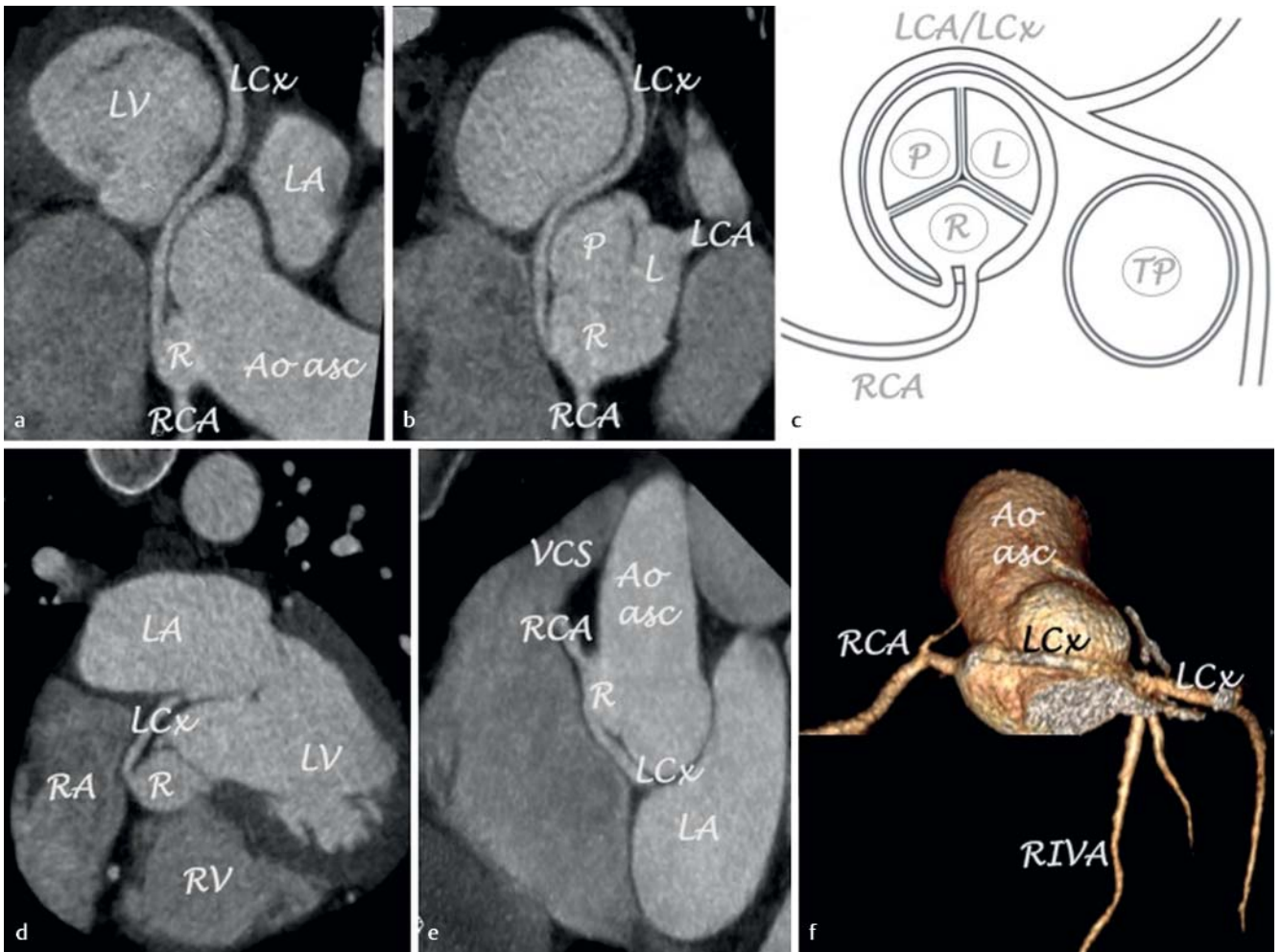
▶ **Fig. 7** Anomalous origin of LCx or LCA from RCS with retroaortic course; **a**, **b** curved reconstruction; **c** scheme; **d** CT-MPR; **e** MIP of curved reconstruction; **f** CT-VRT of coronary artery system; (LA: left atrium; MIP: maximum intensity projection; MPR: multiplanar reconstruction; RA: right atrium; RV: right ventricle; VCS: vena cava superior; further abbreviations Fig. 5).
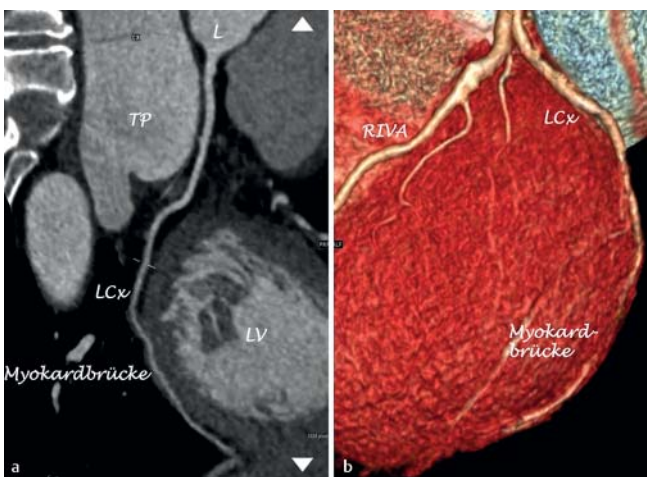
▶ **Fig. 8** Myocardial bridging of the left circumflex artery (LCx); **a** curved reconstruction; **b** CT-VRT; (abbreviations Fig. 5).

▶ **Fig. 9** Coronary artery fistula (CAF) between the left circumflex artery (LCx) and the auricula cordis sinistra (ACS); **a** curved reconstruction; **b** CT-MPR; **c** CT-VRT of coronary artery system; (CAF: coronary artery fistula; ACS: auricula cordis sinistra; abbreviations Fig. 5).

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