Coronary Artery Anomalies

Andrew M. Crean

Contents

1 Introduction

Coronary anomalies are uncommon yet occur not infrequently in the practice of any regular cardiac imager. They can be among both the most challenging and satisfying of lesions to investigate, and frequently noninvasive imaging represents the standard of reference for full description and understanding of the abnormality in question. This article uses a broad definition of coronary anomalies to include all non-atherosclerotic forms of coronary abnormality—not simply congenital anomalies, but also inflammatory anomalies (e.g., due to Kawasaki or Behcet disease) and postsurgical anomalies (e.g., after Ross, Bentall, or arterial switch procedure).

2 Prevalence of Congenital Coronary Anomalies

The prevalence of congenital coronary anomalies appears to have been rising in recent years, and yet this is almost certainly ascertainment bias due to the increased use of cardiac CT in particular which may identify anomalies without great clinical significance (as well as a much smaller number of prognostically important lesions). Most case series have reported a prevalence of around 1% when large cohorts of patients undergoing either coronary angiography or cardiac CT are examined (Table 1).

A.M. Crean, M.R.C.P., F.R.C.R.
Sanghvi Endowed Chair in Cardiovascular Imaging Professor of Cardiology and Pediatrics, University of Cincinnati Medical Center and Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA
e-mail: andrewcrean@gmail.com

Med Radiol Diagn Imaging (2018)
DOI 10.1007/174_2017_170, © Springer International Publishing AG
Published Online: 09 March 2018

Table 1 Prevalence of congenital coronary anomalies in different series

First author	Country	Modality of diagnosis	Population size	Prevalence (%)	Commonest anomaly
Namgung (2014)	South Korea	Cardiac CT	8864	1.16	AORCA
Yuksel (2013)	Turkey	Coronary angiography	16,573	1.3	Separate origin LAD, Cx
Xu (2012)	China	Cardiac CT	12,145	1.02	AORCA
Sohrabi (2012)	Iran	Coronary angiography	6065	1.3	Separate origin LAD, Cx
Sivri (2012)	Turkey	Coronary angiography	12,844	0.74	AOLCx
Fujimoto (2011)	Japan	Cardiac CT	5869	1.52	AORCA
Erol (2011)	Turkey	Cardiac CT	2096	1.96	AORCA and separate origin LAD, Cx
Garg (2000)	India	Coronary angiography	4100	0.95	AORCA
Graidis (2015)	Greece	Cardiac CT	2572	2.33[a]	Separate origin LAD, Cx
Yamanaka (1990)	USA	Coronary angiography	126,595	1.3[a]	Separate origin LAD, Cx
Yildiz (2010)	Turkey	Coronary angiography	12,457	0.9	Separate origin LAD, Cx
Tongut (2016)	Turkey	Cardiac CT	2401	9.37[b]	Separate origin LAD, Cx
Shabestari (2012)	Iran	Cardiac CT	2697	3.1	Separate origin LAD, Cx
Eid (2009)	Lebanon	Coronary angiography	4650	0.73	AOLCx

LAD left anterior descending, *Cx* circumflex, *AORCA* anomalous origin of the right coronary artery (from the opposite sinus), *AOLCx* anomalous origin of the left circumflex

[a]Did not exclude abnormalities of high takeoff which are excluded a priori in most other series

[b]Did not exclude myocardial bridges which are relatively common in the general population—exclusion of these 100 patients reduced their overall prevalence to 5.7%. A further 85 patients were said to have coronary aneurysm or ectasia (no definition provided in methods) which could well have been atherosclerotic (mean age 56 years) or due to prior Kawasaki disease. Exclusion of these patients reduces the prevalence of "true" congenital coronary anomalies to 2.1% which is closer to most other published series

3 Classification of Coronary Anomalies?

Coronary anomalies may initially be subdivided into those present at birth and those which develop at some point after birth because of inflammation or vasculitis (Kawasaki disease, Behcet disease, etc.) or secondary to an operation which involves detachment and reimplantation of the coronary arteries. This chapter is principally concerned with the former, although the latter two categories will be touched upon briefly.

After this basis subdivision, congenital coronary anomalies are best broken down further by the simple classification of:

(a) Abnormalities of origin
(b) Abnormalities of course
(c) Abnormalities of termination

Examples of each are provided in Table 2. These categories are not always totally discrete—abnormalities of origin and proximal course, for example, may accompany one another. Most of the abnormalities in categories a and b are benign, although a few are not. The most significant abnormalities in this regard are the origin of the left or

Table 2 Examples of the most common coronary anomalies and their relative clinical significance (in the author's opinion)

Congenital	Anomaly	Clinical importance
Abnormalities of origin	Anomalous aortic origin from the opposite coronary sinus	High (especially if proximal intramural course)
	Ectopic origin above aortic sinus	Low
Abnormalities of course	Transseptal/infundibular left coronary	Low
	Retro-aortic circumflex	Low
	Pre-pulmonic LAD	Low
	Myocardial bridging	Low
Abnormalities of termination	Circumflex to coronary sinus/right atrial fistula	Intermediate
	Anomalous left coronary artery draining to the pulmonary artery (ALCAPA)	High
	Anomalous right coronary artery draining to the pulmonary artery (ARCAPA)	Intermediate/low
Inflammatory or Vasculitic	Kawasaki disease	High (if aneurysms)
	Behcet disease	High (if aneurysms)
	Takayasu arteritis	High (if stenoses)
Postoperative	Post-arterial switch	Intermediate/low
	Post-Ross procedure	Low
	Post-Bentall procedure	Low

right coronary artery from the opposite sinus of Valsalva. Anomalous origin of the right coronary artery is being seen increasingly frequently due to rapid uptake of cardiac CT and often generates management dilemmas, particularly when a patient presents with symptoms that *might* represent angina but where no objective evidence of ischemia can be found.

Abnormalities of termination generally involve fistulation into another vascular structure or a cardiac chamber. They range from the trivial to the significant.

4 Are All Coronary Anomalies Equally Dangerous?

Table 2 suggests that the answer to this question is no. A more difficult question, however, is *which* anomalies may be life-threatening and how do we identify them. Our knowledge of the natural history of coronary anomaly lesions is incomplete and inadequate. Most of our data is derived from selected pathologic series of civilian deaths or deaths of military recruits during training. In these samples the larger denominator—the numbers of people with identical anomalies but who do *not* die—is unknown.

4.1 Anomalies of Origin

Awareness of the potential for cardiac mortality was raised almost 50 years ago following review of the exhibits at the Armed Forces Institute of Pathology (Cheitlin et al. 1974). This series was among the first to suggest that origin of the left main or left anterior descending (LAD) coronary artery from the right sinus appeared to be disproportionately associated with sudden cardiac death in the setting of exercise. Although not clearly described, the implied mechanism was that of ostial narrowing of the vessel as it emerged obliquely through the aortic wall (Fig. 1). They further suggested that as aortic pressure increases with exercise, there could be a "flap-valve" effect whereby the ostial abnormality

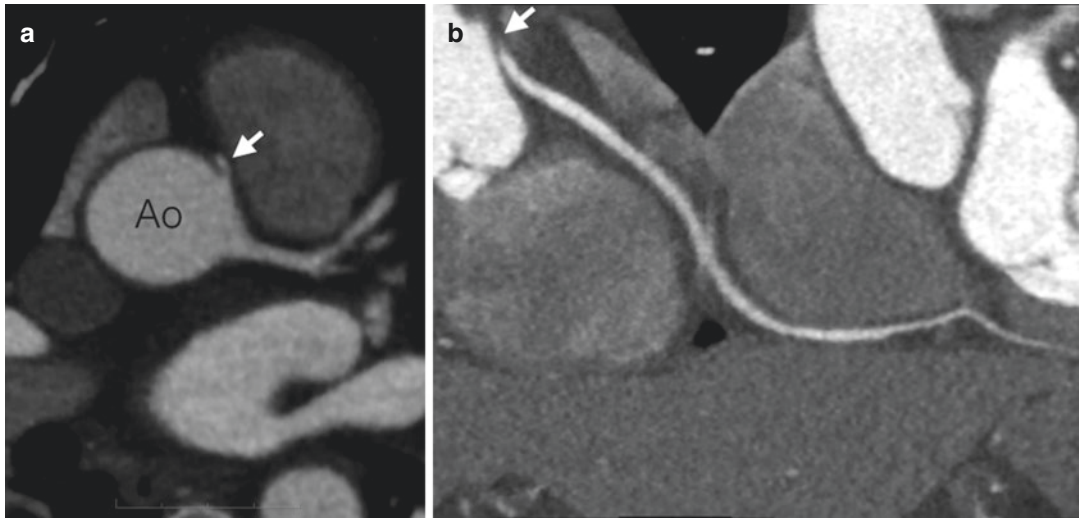

Fig. 1 Intramural right coronary artery (RCA) arising from the left coronary sinus. (**a**) Gated axial CT image at the level of the aortic root (Ao). The RCA arises from the left coronary cusp, and in its most proximal portion, it takes an intramural course through the aortic wall (arrow). (**b**) The effect of intramural passage in creating a slit-like orifice is better appreciated on this curved multiplayer reformat where the ostial portion of the vessel (arrow) appears tightly stenosed compared to its more distal caliber

transiently worsens. Interestingly they saw no cases where an anomalous right coronary artery (RCA) arising from the left sinus of Valsalva led to sudden death; however, it is worth noting that this anomaly—which they display in Fig. 2 of their original publication—does *not* demonstrate the slit-like intramural course which we know to be present from cardiac CT data in this era. Therefore we must question whether their data on this lesion can really be applied to the patient population we see increasingly.

A similar review was performed by the Pathology branch of the National Heart Lung and Blood Institute (Kragel and Roberts 1988). In this series five out of seven cases where the left main (LM) coronary artery arose from the opposite sinus resulted in sudden death. Outcome for patients where the RCA arose from the opposite sinus was more variable with only 8 out of 25 dying as a result of their coronary lesion. In this paper great attention was paid to morphology of the coronary ostia. Out of the 25 cases of RCA from the opposite sinus, 14 were shown to have slit-like morphology of whom only 6 were believed to have died because of the anomaly itself (clear alternative causes of death in the remaining 8 patients). Interestingly, however, two of the patients with RCA from the opposite sinus but *without* a slit-like origin were also classified as having died suddenly and presumably related to their anomaly as no other cause was determined. Overall then, eight patients were presumed to have died from their RCA abnormality; however, the body of the paper goes on to mention that three of these eight had evidence of at least one 75% stenosis in an epicardial coronary vessel and one out of the eight had evidence of hypertrophic cardiomyopathy. So in effect four of the eight sudden deaths may have had an alternative explanation. A further possible confounder is whether or not an anomalous RCA provides the posterior descending artery (i.e., shows right coronary dominance) since this determines the extent of myocardium at risk.

A third necropsy series considered ten patients with RCA from the opposite sinus, three of whom died presumably due to acute ischemia caused by the lesion (each case related to exercise) and seven in whom the abnormality was an incidental finding after death from another cause (Roberts et al. 1982). Interestingly each one of the ten cases had a slit-like RCA ostium, but what

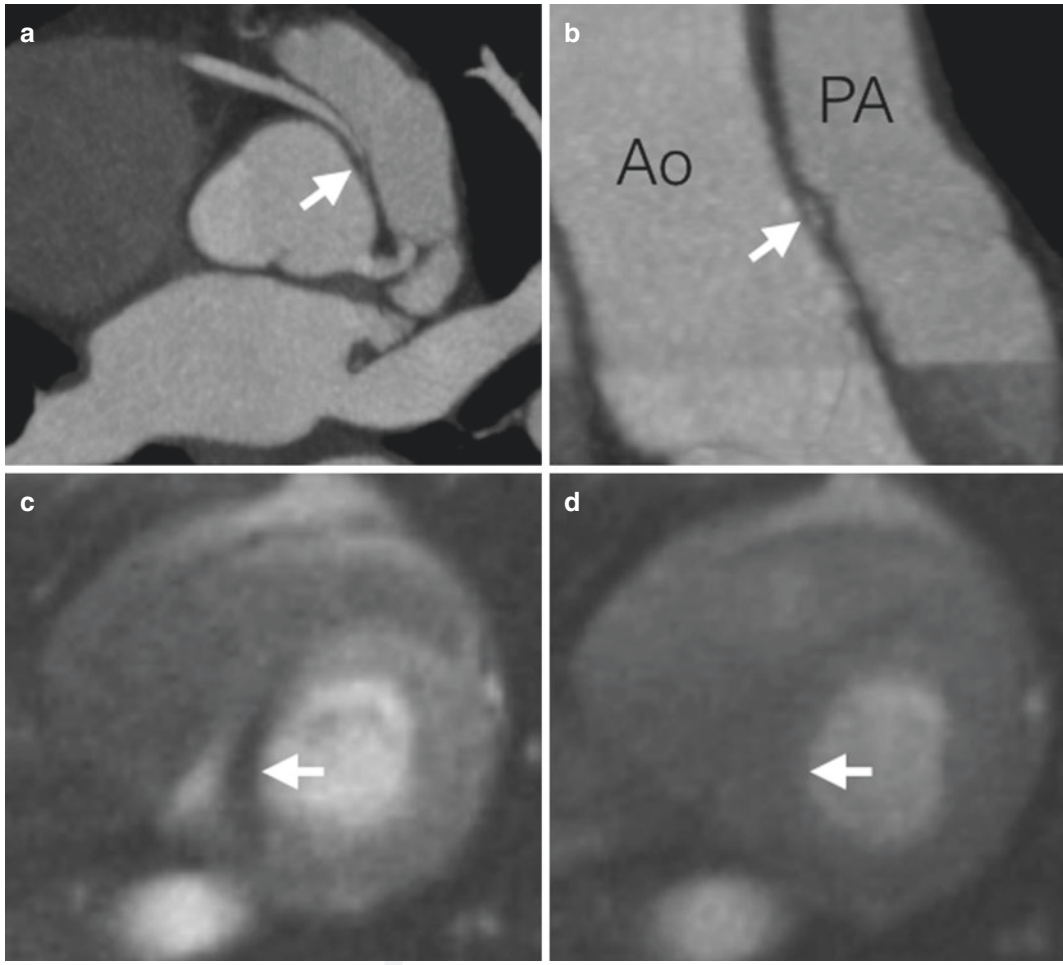

Fig. 2 Anomalous RCA from the left coronary sinus with evidence of ischemia. (**a**) Axial slice from gated CT at the level of the aortic root demonstrates that the RCA arises from the left coronary cusp and has a slit-like origin in its proximal portion (arrow). (**b**) Sagittal oblique reconstruction from gated CT to show the relationship between the aortic root (Ao) and main pulmonary artery (PA). The slit-like ostium can be appreciated in cross section (arrow) as the vessel runs through the aortic wall. (**c**) Basal slice from dipyridamole stress perfusion CMR. A low-signal perfusion defect is present in the inferior septum (arrow). (**d**) Basal slice from dipyridamole stress perfusion CMR 6 months after unroofing surgery which alleviated the patient's symptoms. No evidence of a perfusion abnormality is seen on stress imaging

seemed to be more significant was the presence of macroscopic left ventricular scarring which was present in all three of the sudden death cases but only one out of seven of the noncardiac death cases (and this is in a patient with severe atherosclerotic disease). The same four patients also were the only ones to have evidence of myocardial interstitial fibrosis at histopathology.

At the current time the general consensus is that when the left main or LAD coronary artery arises from the opposite sinus, the patient should be offered surgical repair. Given the rare occurrence of this pathology, no survival or outcome data are available. Anomalous origin of the RCA does not necessarily require repair, particularly when an incidental finding. Patients, however, who have anginal symptoms or evidence of ischemia on testing (Fig. 2) should be considered for repair on a case-by-case basis.

4.2 Anomalies of Termination

This kind of anomaly includes coronary fistulae and anomalous left coronary artery from the pulmonary artery (ALCAPA) which in a way may also considered to be a special kind of fistula. The frequency of congenital coronary fistulae is roughly 0.2% (Gillebert et al. 1986). If fistulae are large in size, they may lead to progressive volume loading of a ventricular chamber with the risk for gradual deterioration in myocardial function (Lee and Chen 2009). Large fistulae may also be associated with a risk of myocardial ischemia (see ALCAPA, below). Another risk which has been associated with fistulae is that of bacterial endocarditis (Said 2016). Finally—although underappreciated—there is a mortality risk associated with long duration fistulae, particularly if untreated (Nakahira et al. 2007; Rajs et al. 2001; Dichtl et al. 2005; Lau 1995; Bartoloni et al. 2012; Lozano et al. 2008). It has been suggested that the risk of rupture associated with fistulae may be higher in women than men (Said et al. 2008) (Fig. 3).

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a particularly important type of fistulous lesion since it can be associated with sudden cardiac death in both children and adults. This is assumed to be secondary to ischemia and arrhythmia occurring as a result of myocardial steal, as coronary flow is exposed to low-pressure runoff into the pulmonary artery which invariably has a lower diastolic pressure than the aorta. Although the majority of such lesions are detected in early childhood, there are regular reports of first detection of ALCAPA in adulthood (Basha et al. 2016; Hofmeyr et al. 2009; Sajjadieh Khajouei et al. 2016; Toumpourleka et al. 2015; Liu and Miller 2012; Lopes et al. 2011) and in rare cases, even in the elderly (Sadanandan et al. 2012; Fierens et al. 2000). As with other kinds of fistula, there may be aneurysmal dilatation of either left or right coronary artery (Bajona et al. 2007), and this can occur even *after* surgical repair (Bravo-Valenzuela and Silva 2015). On occasion the initial presentation in adult life may be with heart failure or cardiac arrest (Safaa et al. 2013; Quah et al. 2014; Krexi and Sheppard 2013; Kristensen et al. 2008; Pachon et al. 2015; Ripley et al. 2014; Raghuram et al. 2004; Kang et al. 2007; Frapier et al. 1999). For a comprehensive review of ALCAPA imaging in adults, see Pena et al. (2009).

5 Structured Imaging of Coronary Anomalies

Patients are generally sent for imaging with varying degrees of prior knowledge about their lesion. In some cases the provided information may be misleading or frankly incorrect. It is important

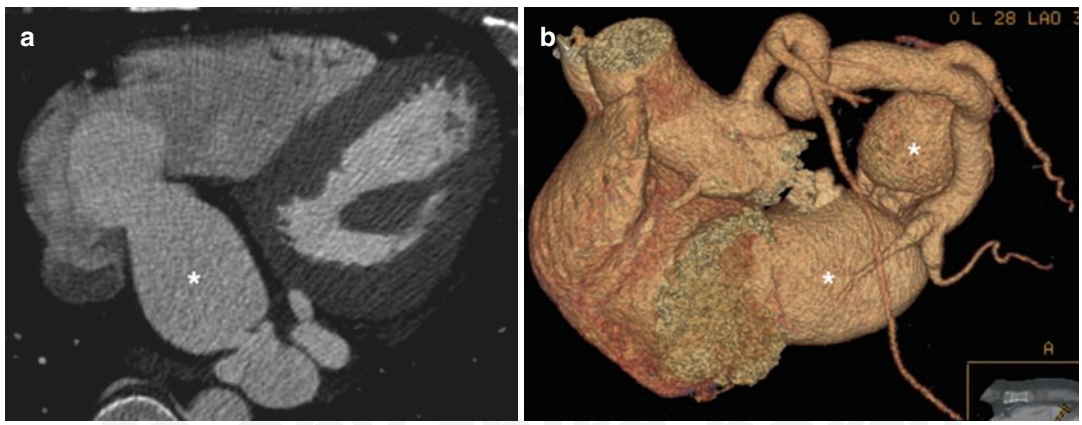

Fig. 3 Left circumflex artery to coronary sinus fistula. (**a**) Gated axial CT image demonstrating a very dilated coronary sinus (asterisk) draining into the right atrium. (**b**) Volume-rendered CT image of the same patient showing more clearly the fistulous communication between the circumflex artery and the coronary sinus

therefore to consider a structured and comprehensive imaging approach for imaging coronary anomalies. The key issues to be addressed by imaging are coronary anatomy, myocardial ischemia, myocardial scar, and the presence or absence of coronary thrombus.

5.1 Anatomic Imaging

Any assessment of a coronary anomaly necessitates a clear understanding of the entire course of the vessel in question—from origin to termination. Although some patients may be sent to the noninvasive lab *after* coronary angiography, invasive angiography is rarely the best method to appreciate the three-dimensional relationship between the coronary artery and surrounding structures. Cardiac MR has been touted as a reliable method for assessing coronary anomalies and certainly can provide a level of information about the vessel's course. However the most frequently used technique for coronary imaging by CMR is a free-breathing navigated steady-state free precession (SSFP) approach which is very dependent upon both heart rate and respiratory pattern for data acquisition. A complete axial 3D volume may therefore take 10–15 min to acquire. Since the quality of the volume stack of images can not be assessed until the last slice is acquired, it unfortunately happens on occasion that the overall image quality is inadequate and up to 10-15 minutes of imaging time will have been wasted acquiring. Nonetheless if a CMR study is planned, we usually attempt coronary imaging and often find it helpful. In situations where the right coronary artery is suspected to arise from the left coronary sinus, or where there is concern for a slit-like origin, then our experience has been that coronary imaging by CMR is often insufficient for diagnostic certainty, and we proceed to cardiac CT in those circumstances.

Cardiac CT is an excellent modality for anatomic coronary imaging, facilitating visualization of coronary origin, course, termination (Su et al. 2010), as well as luminal remodeling, stenosis, and aneurysm. Modern scanners invariably permit prospectively triggered acquisition which limits radiation exposure to a small portion of the cardiac cycle. Overall doses for coronary CT are usually in the range of 1–3 mSv which compares very favorably with invasive angiography and SPECT imaging. In thin patients it is possible to perform sub-millisievert studies through judicious reduction in tube kilovoltage (kV). As a result cardiac CT is now a reasonable option in children (Shen et al. 2016) although historical concerns regarding dose have limited its usage (which is ironic considering how widespread the use of SPECT imaging is in many pediatric institutions).

The isotropic spatial resolution of a modern CT scanner is in the region of 0.5 mm^3, which is 2–3 times better than that achieved by standard CMR angiography and is the reason why the potential proximal intramural portion of a coronary anomaly is better assessed by CT than CMR. The other benefit of CT is its ability to detect atherosclerotic coronary disease which—in patients who present at an older age—may be the real reason for symptoms rather than the coronary anomaly, accused but an innocent bystander (Keir et al. 2017; Crean et al. 2008).

5.2 Ischemia Imaging

The single biggest concern for most anomalies is whether or not they have the potential to result in myocardial ischemia and, subsequently, arrhythmic sudden cardiac death. There is no consensus in the literature or among experts as to how coronary anomalies should be investigated—as a result most groups work toward their own internal consensus (Keir et al. 2015; Gräni et al. 2017). In many centers, myocardial perfusion stress/rest SPECT imaging is utilized to stratify risk. However the larger cohort studies all contain individuals who dropped dead at some time point after a previously normal perfusion study which is hardly reassuring (Basso et al. 2000). More pertinent perhaps is the frequency of false positive results, both before and *after* surgical repair (Brothers et al. 2007).

Stress echo with exercise or dobutamine for wall motion assessment may be a reasonable choice for the assessment of ischemia given its

widespread availability. Myocardial contrast perfusion echo on the other hand is much less practiced, and its use is only very rarely described in the setting of coronary anomalies (Rana et al. 2009). As with all other forms of noninvasive imaging, evidence supporting the diagnostic and prognostic power of stress echo is lacking in this population (Thompson 2015).

Stress perfusion CMR has been shown to have a substantially higher diagnostic *and* prognostic accuracy than SPECT imaging for obstructive atheromatous coronary artery disease (Greenwood et al. 2012, 2016). Conflicting nuclear data suggest that triage by either technique is reasonable with similar medium-term outcomes (Sharples et al. 2007; Thom et al. 2014). There are few corresponding data for congenital heart disease patients, although one study which directly compared isotope perfusion to CMR perfusion in an adult arterial switch population demonstrated a high false positive rate for the nuclear technique (Tobler et al. 2014). Coronary stenosis after coronary reimplantation is relatively rare but is seen on occasion (Fig. 4). We have now studied over 100 patients with stress perfusion CMR in the Toronto Coronary Anomaly Clinic, and our experience is described elsewhere (Deva et al. 2014; Crean et al. 2016). Briefly, among those where a formal reference standard was available for comparison, stress perfusion cardiovascular magnetic resonance demonstrated a sensitivity of 82% and specificity of 100%. Of the 34 studies, two were false negatives, in which the etiology of ischemia was extrinsic arterial compression rather than intrinsic coronary luminal narrowing.

We and others have found that the ALCAPA lesions invariably show evidence of myocardial perfusion abnormality (Nony et al. 1992) but that anomalies of coronary origin—particularly the right coronary from the left cusp with proximal intramural course—are much more variable. Our approach has been to seek to confirm either a positive or negative vasodilator stress CMR study with a second modality, usually stress echocardiography. This latter is chosen because of persistent concerns in some quarters that physiologic stress which raises the blood pressure and dilates the aortic root may promote ischemia to a greater extent than simple drug-induced coronary vasodilatation.

Any anomaly which is demonstrated to generate ischemia is usually subjected to surgical repair. The more difficult scenario arises when stress CMR and stress echo are both negative but the patient continues to complain of symptoms. In this situation we take an aggressive approach with coronary catheterization combined with intravascular ultrasound (IVUS) and measurement of fractional flow reserve (FFR). A 50% reduction in vessel cross-sectional origin at the ostium or an FFR < 0.8 generally results in a decision to proceed with surgical repair. If both

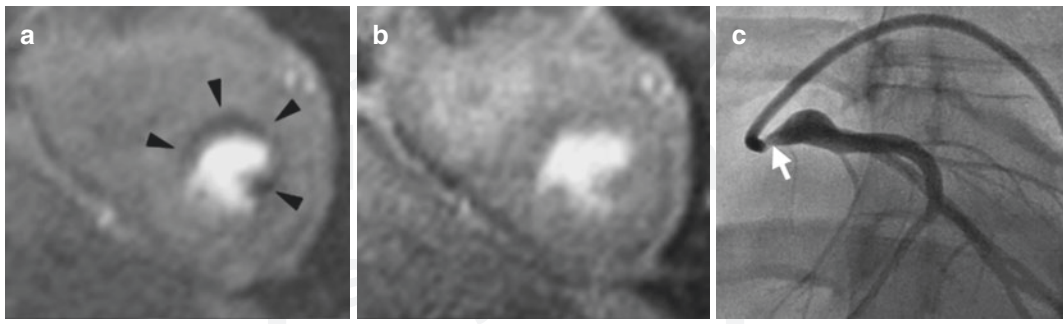

Fig. 4 Ischemia as a long-term complication of the arterial switch procedure. (**a**) Still frame from dipyridamole stress perfusion CMR study demonstrating subendocardial ischemia (arrowheads) in the left coronary territory. (**b**) Rest perfusion image shows normal perfusion. (**c**) Invasive angiogram depicting a significant left main stem coronary artery stenosis (arrow)

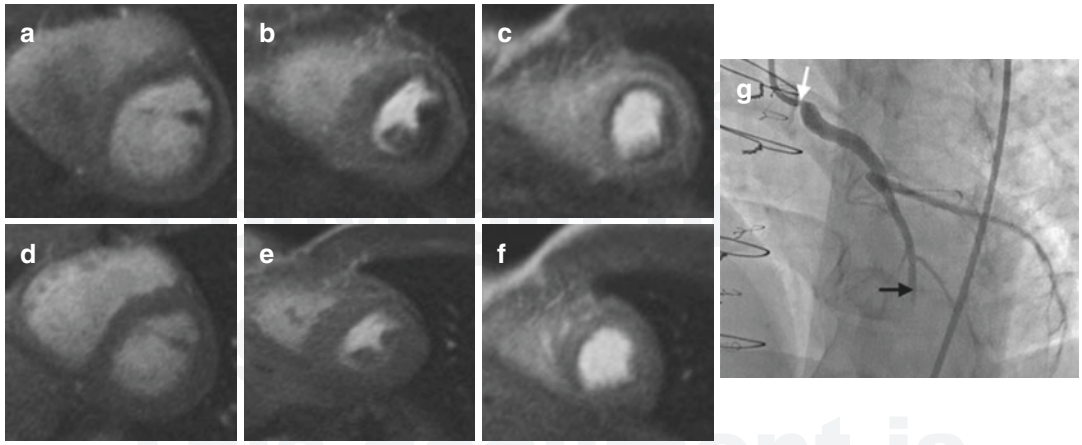

Fig. 5 Tight stenosis of reconstructed left main coronary artery following ALCAPA repair. (**a**–**c**) Basal, mid, and apical short axis slices from dipyridamole stress CMR study. There is an extensive low-signal defect throughout the left coronary territory. (**d–f**) The defect appears full reversible in matching locations at rest. (**g**) Invasive angiogram several days later. There is an extremely tight focal stenosis at the site of implantation of the left main coronary (arrow). Note also incomplete LAD filling due to competitive retrograde flow (black arrow) which still exists from the right coronary collateral network. The patient subsequently underwent left main stem stenting

IVUS and vasodilator FFR are normal, it may be reasonable to repeat FFR measurements with full dobutamine stress to mimic exercise (Angelini and Flamm 2007; Angelini 2014; Angelini et al. 2015; Lee et al. 2016).

Consideration should always be given to an early postoperative CMR study in order to establish a new baseline appearance should symptoms recur in the future. This is particularly important for ALCAPA patients in whom perfusion may not entirely normalize post repair (Seguchi et al. 1990) or where occlusion at the reimplantation site is suspected (Secinaro et al. 2011) (Fig. 5).

5.3 Scar Imaging

Patients with coronary anomalies are at risk of coronary thrombosis or, occasionally, coronary thromboembolism—both potentially resulting in myocardial infarction. There is little doubt that CMR is the gold standard for detection of myocardial scar, and late gadolinium enhancement (LGE) should be included in every CMR protocol when assessing coronary anomalies (Fig. 6). In patients with prior cardiac surgery (e.g., Ross, Bentall, or arterial switch procedure), it should be borne in mind that scar is not always due to the anomaly itself and that prior cardiopulmonary bypass may result in small areas of scarring. Patients who have had VSD patch closure may demonstrate an LGE signal in the region of repair due to overlying fibrotic tissue, and this needs to be distinguished from true myocardial damage. Genuine scar is a risk factor for adverse remodeling and arrhythmogenesis as it is in the noncongenital population and ideally should be quantitated in the imaging report. There is also increasing research interest in subclinical fibrosis in congenital cardiology using techniques such as T1 and ECV mapping (de Meester de Ravenstein et al. 2015; Burchill et al. 2017).

A small number of patients are unable to undergo LGE imaging due to claustrophobia or other CMR contraindications. In these cases SPECT imaging may substitute although it is insensitive for small amounts of scar (Wagner et al. 2003). However, if cardiac CT is planned, then a second study some minutes after contrast injection may be considered to demonstrate infarction by the technique of late iodine enhancement (Kramer et al. 1984) which is

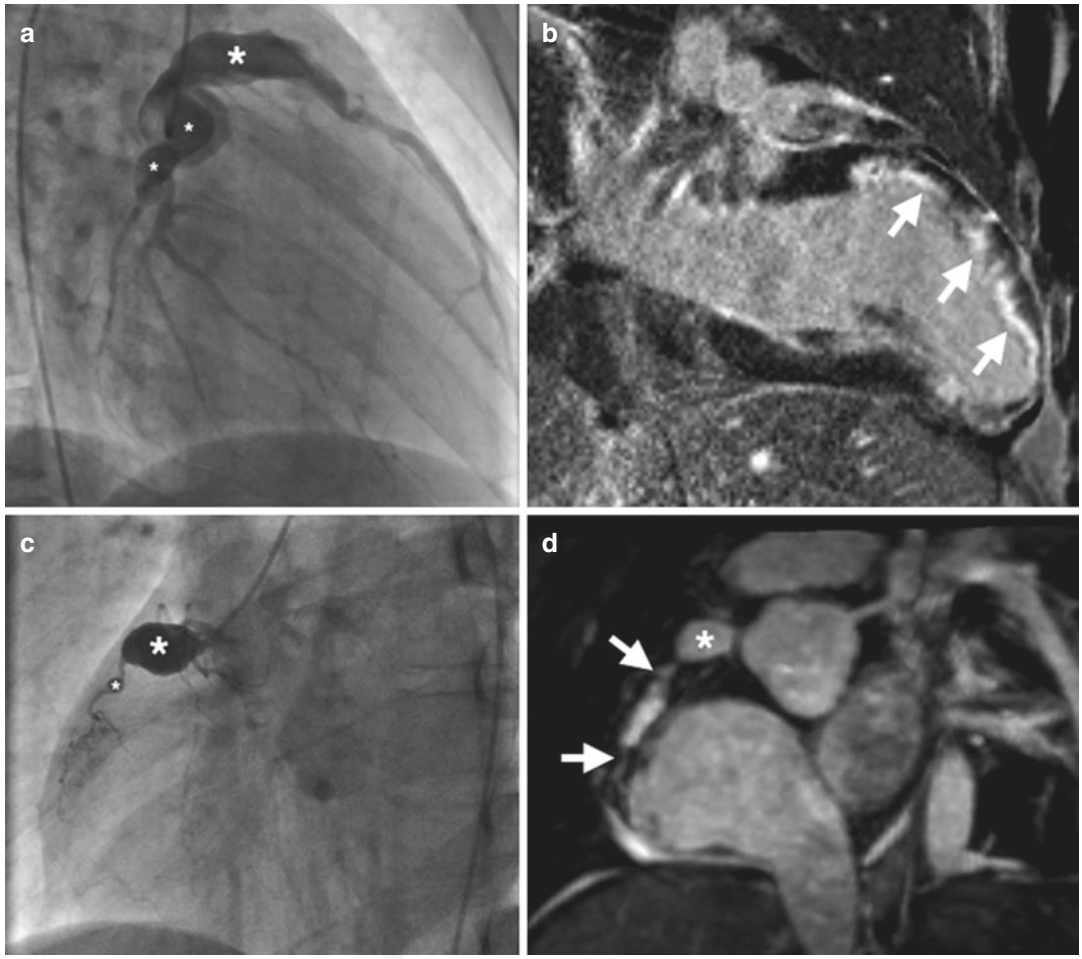

Fig. 6 Kawasaki disease complicated by myocardial infarction. (**a**) Invasive angiogram with injection into the left coronary system. There is a large tubular LAD aneurysm (asterisk) and two smaller sequential circumflex artery aneurysms (smaller asterisks). (**b**) Two chamber late gadolinium enhancement CMR image demonstrating an extensive subendocardial infarct (arrows) in the LAD territory. This occurred in adolescence in association with acute aneurysm thrombosis after the patient self-discontinued anticoagulation therapy. (**c**) Invasive angiogram in the same patient shows a large proximal RCA aneurysm (large asterisk) together with a much smaller one more distally (smaller asterisk). (**d**) Short axis reformat from whole-heart navigated free-breathing CMR coronary angiogram. The aneurysms in the RCA are clearly appreciated, but note that delineation of the intervening vessel segments (arrows) is inferior compared to invasive angiography (prior frame)

analogous to LGE imaging albeit with lower contrast and signal to noise (Jacquier et al. 2008). This has been validated in animals (Baks et al. 2006; Buecker et al. 2005; Burk et al. 2015; Lardo et al. 2006; Crean et al. 2013) and in humans by CMR (Deux et al. 2013; Habis et al. 2009), electroanatomic mapping (Esposito et al. 2016), and positron emission tomography (Dwivedi et al. 2013). The technique is less robust than LGE imaging, and neither the optimal dose of contrast nor the delay prior to image acquisition is well validated. Anecdotal reports indicate that intracoronary injection of contrast may help visualization of thin rims of scar (Baks et al. 2006; Buecker et al. 2005; Burk et al. 2015; Lardo et al. 2006; Crean et al. 2013), but this seems insufficiently pragmatic to become a mainstream approach.

5.4 Thrombus Imaging

The risks of thrombus formation relate to perturbations of Virchow's triad as much for a coronary anomaly as anywhere else in the cardiovascular system. Thus all three aspects—altered vessel, altered flow, altered coagulability—need to be considered (Lowe 2003). Vascular and flow aspects can be determined by CMR/CT, and knowledge of the patient's coagulation status (medications, pregnancy, smoking, obesity, cancer, etc.) is valuable when interpreting these studies.

Thrombus is only rarely a consideration in large fistulae where flow is brisk. However the circumflex artery to coronary sinus fistula has been associated with myocardial infarction (Dichtl et al. 2005; Al-Turki et al. 2015). In our clinic we have seen one such case arising in a young woman after several weeks of immobilization post orthopedic surgery, presumably reflecting reduced flow due to a much lower basal cardiac output than usual.

The most concerning coronary anomaly where thrombus should be actively excluded is in the setting of Kawasaki disease with giant coronary aneurysms (Patil et al. 2008; El-Segaier and Galal 2013; Ghosh and Agarwala 2011; Mendiola Ramírez et al. 2011; Okura et al. 2013). Aneurysm formation leads to abnormal shear stress and abnormal flow patterns within the ectatic vessel components (Kuramochi et al. 2000). Here there is a real risk of thrombus formation which may lead to distal embolization or acute closure of the vessel (Rizk et al. 2015; Argo et al. 2016; Teo and Paul 2005). Acute Kawasaki disease in children is a diffuse inflammatory state, and care should be taken to exclude thrombi elsewhere within the heart also (Song et al. 2015). Recognition of thrombus is important as aggressive therapy with warfarinization or even thrombolysis needs to be considered (Harada et al. 2013).

Thrombus may be recognized in children by 2D echocardiography as an echogenic mass within the coronary lumen. However in adults cross-sectional imaging is usually required. Both cardiac CT and CMR are believed to be accurate for this purpose, but regardless of which modality is chosen, it is usually necessary to adopt a late enhancement strategy to maximize the low attenuation/signal of the thrombus against the luminal background (Fig. 7). In practical terms this means either using LGE imaging after a standard 10 min delay or alternatively acquiring gated CT images roughly 60–90 s after either a bolus or infusion of iodine (Figs. 8 and 9). CT images acquired in the standard coronary arterial phase (used for routine atherosclerotic CT coronary angiography) may be unreliable as swirling flow within the larger aneurysms often leads to a mixture of opacified and unopacified blood within the aneurysm in the early arterial phase—with the unopacified portion mimicking thrombus.

5.5 Flow Imaging

Flow assessment is generally not a major part of coronary anomaly imaging although on occasion it may be used to help ascertain the point of termination of a fistula when the anatomy is unclear. Flow imaging in order to assess shunt size is important primarily in the setting of coronary fistula. Phase-contrast CMR is used to measure both aortic and pulmonary flow for calculation of shunt ratio—large shunts will often be associated with chamber enlargement, and this in itself is often an adequate reason for surgical repair. Shunt ratio may also be formally estimated by cardiac catheterization, and there is reasonably good correlation between this modality and CMR estimation of shunt (Hundley et al. 1995).

In the specific setting of ALCAPA/ARCAPA, the dilated collateral network between the right and left coronary systems gives rise to a number of echocardiographic features (Courand et al. 2013; Silverman 2015; Yang et al. 2007) including systolic coronary flow predominance which has not been reported in any other coronary anomaly (Ghaderi et al. 2014). Three-dimensional echo may be useful intraoperatively for demonstrating the site of connection of the left coronary artery to the surgeon in real time (Jin et al. 2011).

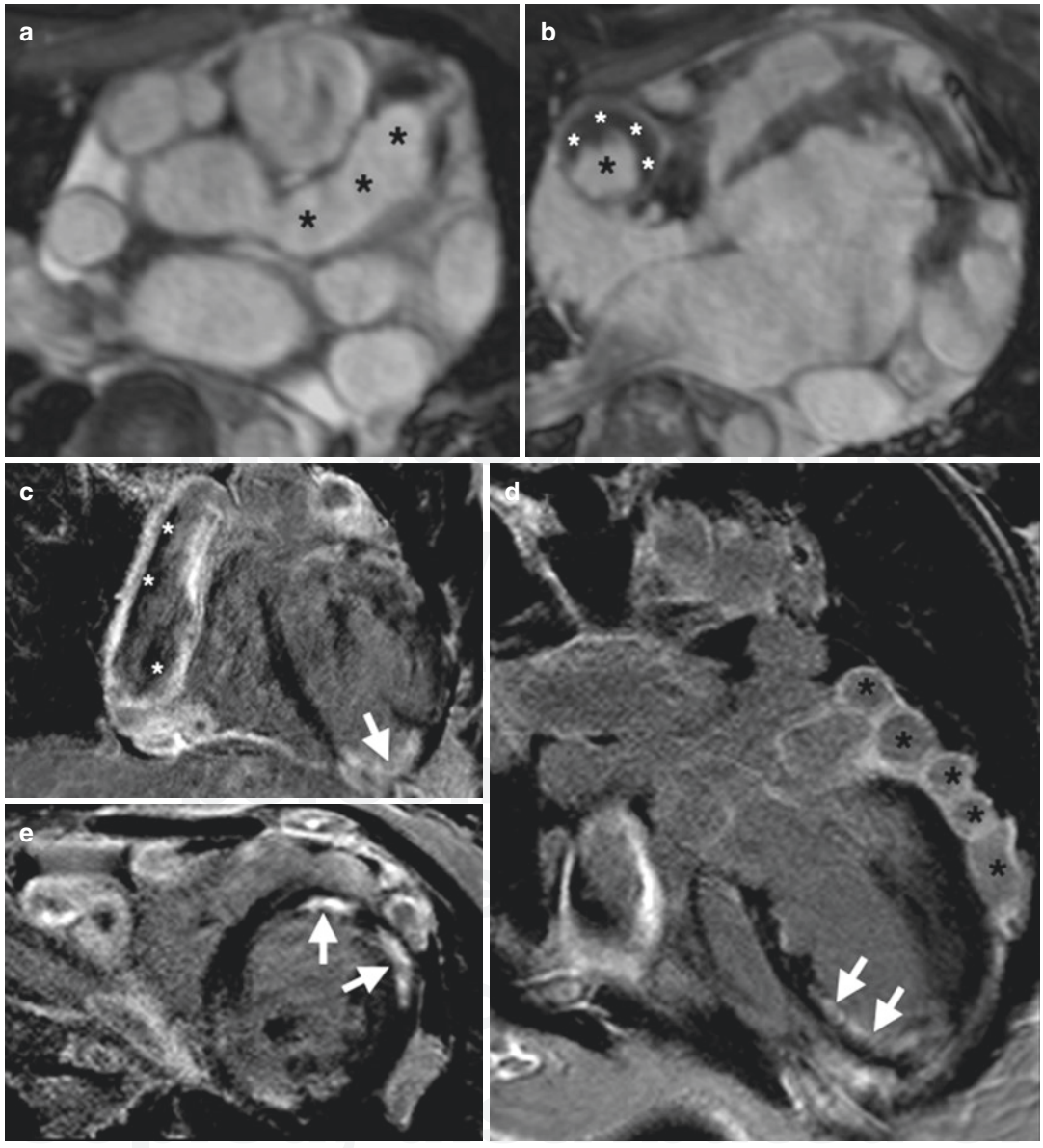

Fig. 7 Noonan syndrome with diffuse severe coronary ectasia, thrombus, and embolic infarction. (**a**) Axial image from whole-heart coronary CMR angiogram. There is severe ectasia of the left main coronary artery (asterisks) up to around 15 mm (normal <5 mm). (**b**) CMR whole-heart angiogram demonstrating a rim of thrombus (white asterisks) in a severely dilated right coronary artery (black asterisk). (**c**) Coronal late gadolinium enhancement image demonstrates low-signal mural thrombus (asterisks) in the RCA which measures up to 20 mm in diameter (normal 2–4 mm). (**d**) Four-chamber LGE image demonstrates subendocardial infarction (arrows) in the mid to distal septum. In addition the bizarrely dilated circumflex artery is seen as a "string of beads" along the left lateral aspect of the ventricle (black asterisks). (**e**) Short axis LGE view demonstrates additional scar (arrows) in the mid-anterior wall

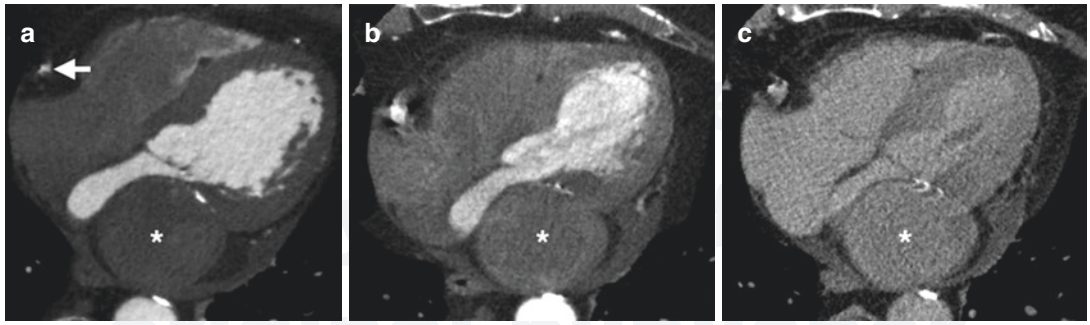

Fig. 8 Cardiac CT in a Kawasaki patient with a giant circumflex artery aneurysm. (**a**) Coronary arterial phase—there is no enhancement of the circumflex artery aneurysm (asterisk) although note that contrast has already enhanced the right coronary artery (arrow). (**b**) Late arterial phase—uneven attenuation is present throughout the aneurysm which could be due to the presence of thrombus. (**c**) Extended venous phase—these images were acquired at 6 min from injection, and this was the first time point at which uniform enhancement was seen throughout the aneurysm. There is no evidence of thrombus (but see Fig. 9)

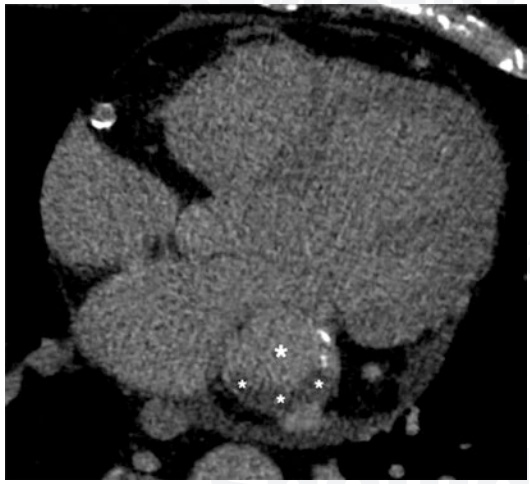

Fig. 9 Cardiac CT demonstrating thrombus in a Kawasaki aneurysm. Same patient as Fig. 8 but at a different place in the circumflex artery. The images have been acquired in a late venous phase at 6 min after contrast, although it is unusual to have to wait this long. The aneurysm (larger asterisk) is uniformly enhanced except for a posterior low attenuation crescentic rim (smaller asterisks) which represents chronic mural thrombus

6 Summary

Coronary imaging is often required for non-atherosclerotic congenital anomalies as well as for inflammatory/vasculitic conditions and in the setting of postoperative coronary repair. No single form of imaging is universally applicable, and complicated cases are likely to require a multimodality approach. Every attempt should be made to demonstrate ischemia in order to justify surgery. On rare occasions, however, there may still be circumstances where surgery can be justified even in the absence of a positive study. Close collaboration between imagers, cardiologists, and cardiac surgeons is vitally important in these cases.

References

Al-Turki MA, Patton D, Crean AM, Horlick E, Dhillon R, Johri AM (2015) Spontaneous thrombosis of a left circumflex artery fistula draining into the coronary sinus. World J Pediatr Congenit Heart Surg 6(4):640–642

Angelini P (2014) Novel imaging of coronary artery anomalies to assess their prevalence, the causes of clinical symptoms, and the risk of sudden cardiac death. Circ Cardiovasc Imaging 7(4):747–754

Angelini P, Flamm SD (2007) Newer concepts for imaging anomalous aortic origin of the coronary arteries in adults. Catheter Cardiovasc Interv 69(7):942–954

Angelini P, Uribe C, Monge J, Tobis JM, Elayda MA, Willerson JT (2015) Origin of the right coronary artery from the opposite sinus of Valsalva in adults: characterization by intravascular ultrasonography at baseline and after stent angioplasty. Catheter Cardiovasc Interv 86(2):199–208

Argo A, Zerbo S, Maresi EG, Rizzo AM, Sortino C, Grassedonio E et al (2016) Utility of post mortem MRI in definition of thrombus in aneurismatic coronary arteries due to incomplete Kawasaki disease in infants. J Forens Radiol Imag 7:17–20

Bajona P, Maselli D, Dore R, Minzioni G (2007) Anomalous origin of the left main artery from the

pulmonary artery: adult presentation with systemic collateral supply and giant right coronary artery aneurysm. J Thorac Cardiovasc Surg 134(2):518–520

Baks T, Cademartiri F, Moelker AD, Weustink AC, van Geuns R-J, Mollet NR et al (2006) Multislice computed tomography and magnetic resonance imaging for the assessment of reperfused acute myocardial infarction. J Am Coll Cardiol 48(1):144–152

Bartoloni G, Giorlandino A, Calafiore AM, Caltabiano R, Cosentino S, Algieri G et al (2012) Multiple coronary artery-left ventricular fistulas causing sudden death in a young woman. Hum Pathol 43(9):1520–1523

Basha KMS, Vaijyanath P, Thomas S (2016) Adult type ALCAPA–A rare presentation. J Indian Colleg Cardiol

Basso C, Maron BJ, Corrado D, Thiene G (2000) Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. J Am Coll Cardiol 35(6):1493–1501

Bravo-Valenzuela NJM, Silva GRN (2015) Aneurysm of the left coronary artery in postoperative bland-white-garland syndrome. Case Rep Cardiol 2015:568014

Brothers JA, McBride MG, Seliem MA, Marino BS, Tomlinson RS, Pampaloni MH et al (2007) Evaluation of myocardial ischemia after surgical repair of anomalous aortic origin of a coronary artery in a series of pediatric patients. J Am Coll Cardiol 50(21):2078–2082

Buecker A, Katoh M, Krombach GA, Spuentrup E, Bruners P, Günther RW et al (2005) A feasibility study of contrast enhancement of acute myocardial infarction in multislice computed tomography: comparison with magnetic resonance imaging and gross morphology in pigs. Investig Radiol 40(11):700–704

Burchill L, Huang J, Tretter J, Khan A, Crean A, Veldtman G et al (2017) Noninvasive imaging in adult congenital heart disease. Circ Res 120:995–1014

Burk LM, Wang K-H, Wait JM, Kang E, Willis M, Lu J et al (2015) Delayed contrast enhancement imaging of a murine model for ischemia reperfusion with carbon nanotube micro-CT. PLoS One 10(1):e0115607

Cheitlin MD, De Castro CM, McAllister HA (1974) Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva, a not-so-minor congenital anomaly. Circulation 50(4):780–787

Courand P-Y, Bozio A, Ninet J, Henaine R, Veyrier M, Bakloul M et al (2013) Focus on echocardiographic and Doppler analysis of coronary artery abnormal origin from the pulmonary trunk with mild myocardial dysfunction. Echocardiography 30(7):829–836

Crean AM, Kilcullen N, Younger JF (2008) Arrhythmic acute coronary syndrome and anomalous left main stem artery: culprit or innocent bystander. Acute Card Care 10(1):60–61

Crean AM, Spears DA, Suszko AM, Chauhan VS (2013) High-resolution 3D scar imaging using a novel late iodine enhancement multidetector CT protocol to guide ventricular tachycardia catheter ablation. J Cardiovasc Electrophysiol 24(6):708–710

Crean AM, Deva DP, Wald R (2016) Emerging role of stress perfusion cardiovascular magnetic resonance in the patient with congenital heart disease. In: da Cruz EM, Ivy D, Hraska V, Jaggers J (eds) Pediatric and congenital cardiology, cardiac surgery and intensive care. Springer, London, pp 1–22

Deux J-F, Potet J, Lim P, Teiger E, Mayer J, Bensaid A et al (2013) Is multidetector computed tomography a suitable alternative to MR imaging for the assessment of myocardial necrosis after alcohol septal ablation? Int J Cardiol 164(3):306–311

Deva DP, Torres FS, Wald RM, Roche SL, Jimenez-Juan L, Oechslin EN et al (2014) The value of stress perfusion cardiovascular magnetic resonance imaging for patients referred from the adult congenital heart disease clinic: 5-year experience at the Toronto General Hospital. Cardiol Young 24(5):822–830

Dichtl W, Waldenberger P, Pachinger O, Müller S (2005) An uncommon coronary artery fistula causing survived sudden cardiac death in a young woman. Int J Card Imaging 21(4):387–390

Dwivedi G, Al-Shehri H, deKemp RA, Ali I, Alghamdi AA, Klein R et al (2013) Scar imaging using multislice computed tomography versus metabolic imaging by F-18 FDG positron emission tomography: a pilot study. Int J Cardiol 168(2):739–745

Eid AH, Itani Z, Al-Tannir M, Sayegh S, Samaha A (2009) Primary congenital anomalies of the coronary arteries and relation to atherosclerosis: an angiographic study in Lebanon. J Cardiothorac Surg 4:58

El-Segaier M, Galal M (2013) Intracoronary thrombus in an infant with Kawasaki disease and giant coronary aneurysm. Acta Paediatr 102(5):e227–e228

Erol C, Seker M (2011) Coronary artery anomalies: the prevalence of origination, course, and termination anomalies of coronary arteries detected by 64-detector computed tomography coronary angiography. J Comput Assist Tomogr 35(5):618–624

Esposito A, Palmisano A, Antunes S, Maccabelli G, Colantoni C, Rancoita PMV et al (2016) Cardiac CT with delayed enhancement in the characterization of ventricular tachycardia structural substrate: relationship between CT-segmented scar and electro-anatomic mapping. JACC Cardiovasc Imaging 9(7):822–832

Fierens C, Budts W, Denef B, Van De Werf F (2000) A 72 year old woman with ALCAPA. Heart 83(1):E2

Frapier JM, Leclercq F, Bodino M, Chaptal PA (1999) Malignant ventricular arrhythmias revealing anomalous origin of the left coronary artery from the pulmonary artery in two adults. Eur J Cardiothorac Surg 15(4):539–541

Fujimoto S, Kondo T, Orihara T, Sugiyama J, Kondo M, Kodama T et al (2011) Prevalence of anomalous origin of coronary artery detected by multi-detector computed tomography at one center. J Cardiol 57(1):69–76

Garg N, Tewari S, Kapoor A, Gupta DK, Sinha N (2000) Primary congenital anomalies of the coronary arteries: a coronary: arteriographic study. Int J Cardiol 74(1):39–46

Ghaderi F, Gholoobi A, Moeinipour A (2014) Unique echocardiographic markers of anomalous origin of the left coronary artery from the pulmonary

artery (ALCAPA) in the adult. Echocardiography 31(1):E13–E15

Ghosh A, Agarwala BN (2011) Kawasaki disease: giant aneurysm with a large thrombus of the left coronary artery. Clin Pract 1(2):e23

Gillebert C, Van Hoof R, Van de Werf F, Piessens J, De Geest H (1986) Coronary artery fistulas in an adult population. Eur Heart J 7(5):437–443

Graidis C, Dimitriadis D, Karasavvidis V, Dimitriadis G, Argyropoulou E, Economou F et al (2015) Prevalence and characteristics of coronary artery anomalies in an adult population undergoing multidetector-row computed tomography for the evaluation of coronary artery disease. BMC Cardiovasc Disord 15:112

Gräni C, Buechel RR, Kaufmann PA, Kwong RY (2017) Multimodality imaging in individuals with anomalous coronary arteries. JACC Cardiovasc Imaging 10(4):471–481

Greenwood JP, Maredia N, Younger JF, Brown JM, Nixon J, Everett CC et al (2012) Cardiovascular magnetic resonance and single-photon emission computed tomography for diagnosis of coronary heart disease (CE-MARC): a prospective trial. Lancet 379(9814):453–460

Greenwood JP, Herzog BA, Brown JM, Everett CC, Nixon J, Bijsterveld P et al (2016) Prognostic value of cardiovascular magnetic resonance and single-photon emission computed tomography in suspected coronary heart disease: long-term follow-up of a prospective, diagnostic accuracy cohort study. Ann Intern Med 165(1):1–9

Habis M, Capderou A, Sigal-Cinqualbre A, Ghostine S, Rahal S, Riou JY et al (2009) Comparison of delayed enhancement patterns on multislice computed tomography immediately after coronary angiography and cardiac magnetic resonance imaging in acute myocardial infarction. Heart 95(8):624–629

Harada M, Akimoto K, Ogawa S, Kato H, Nakamura Y, Hamaoka K et al (2013) National Japanese survey of thrombolytic therapy selection for coronary aneurysm: intracoronary thrombolysis or intravenous coronary thrombolysis in patients with Kawasaki disease. Pediatr Int 55(6):690–695

Hofmeyr L, Moolman J, Brice E, Weich H (2009) An unusual presentation of an anomalous left coronary artery arising from the pulmonary artery (ALCAPA) in an adult: anterior papillary muscle rupture causing severe mitral regurgitation. Echocardiography 26(4):474–477

Hundley WG, Li HF, Lange RA, Pfeifer DP, Meshack BM, Willard JE et al (1995) Assessment of left-to-right intracardiac shunting by velocity-encoded, phase-difference magnetic resonance imaging. A comparison with oximetric and indicator dilution techniques. Circulation 91(12):2955–2960

Jacquier A, Revel D, Saeed M (2008) MDCT of the myocardium: a new contribution to ischemic heart disease. Acad Radiol 15(4):477–487

Jin YD, Hsiung MC, Tsai SK, Chang C-Y, Wei J, Ou C et al (2011) Successful intraoperative identification of an anomalous origin of the left coronary artery from the pulmonary artery using real time three-dimensional transesophageal echocardiography. Echocardiography 28(7):E149–E151

Kang WC, Chung W-J, Choi CH, Park KY, Jeong MJ, Ahn TH et al (2007) A rare case of anomalous left coronary artery from the pulmonary artery (ALCAPA) presenting congestive heart failure in an adult. Int J Cardiol 115(2):e63–e67

Keir M, Wald RM, Roche SL, Oechslin EN, Horlick E, Osten MD et al (2015) Does a dedicated subspecialty ACHD coronary clinic result in greater consistency in approach and reduced loss to follow-up? An evaluation of the first 3years of the Toronto congenital coronary Clinic for Adults. Prog Pediatr Cardiol 39(2):145–150

Keir M, Spears D, Caldarone C, Crean AM (2017) Proving the innocence of a "malignant" coronary artery: calling dobutamine stress CT for the defence. J Cardiovasc Comput Tomogr 11(1):68–69

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(10 Pt 1):771–777

Kramer PH, Goldstein JA, Herkens RJ, Lipton MJ, Brundage BH (1984) Imaging of acute myocardial infarction in man with contrast-enhanced computed transmission tomography. Am Heart J 108(6):1514–1523

Krexi L, Sheppard MN (2013) Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), a forgotten congenital cause of sudden death in the adult. Cardiovasc Pathol 22(4):294–297

Kristensen T, Kofoed KF, Helqvist S, Helvind M, Søndergaard L (2008) Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) presenting with ventricular fibrillation in an adult: a case report. J Cardiothorac Surg 3:33

Kuramochi Y, Ohkubo T, Takechi N, Fukumi D, Uchikoba Y, Ogawa S (2000) Hemodynamic factors of thrombus formation in coronary aneurysms associated with Kawasaki disease. Pediatr Int 42(5):470–475

Lardo AC, Cordeiro MAS, Silva C, Amado LC, George RT, Saliaris AP et al (2006) Contrast-enhanced multidetector computed tomography viability imaging after myocardial infarction: characterization of myocyte death, microvascular obstruction, and chronic scar. Circulation 113(3):394–404

Lau G (1995) Sudden death arising from a congenital coronary artery fistula. Forensic Sci Int 73(2):125–130

Lee M-L, Chen M (2009) Diagnosis and management of congenital coronary arteriovenous fistula in the pediatric patients presenting congestive heart failure and myocardial ischemia. Yonsei Med J 50(1):95–104

Lee SE, CW Y, Park K, Park KW, Suh J-W, Cho Y-S et al (2016) Physiological and clinical relevance of anomalous right coronary artery originating from left sinus of Valsalva in adults. Heart 102(2):114–119

Liu Y, Miller BW (2012) ALCAPA presents in an adult with exercise intolerance but preserved cardiac function. Case Rep Cardiol 2012:471759

Lopes R, Almeida PB, Amorim MJ, Magalhães D, Silva JC, Pinho P et al (2011 Aug) Anomalous origin of the left coronary artery from the pulmonary artery in an asymptomatic adult. Congenit Heart Dis 6(4):366–369

Lowe GDO (2003) Virchow's triad revisited: abnormal flow. Pathophysiol Haemost Thromb 33(5–6):455–457

Lozano I, Batalla A, Rubin J, Avanzas P, Martin M, Moris C (2008) Sudden death in a patient with multiple left anterior descending coronary artery fistulas to the left ventricle. Int J Cardiol 125(3):e37–e39

de Meester de Ravenstein C, Bouzin C, Lazam S, Boulif J, Amzulescu M, Melchior J et al (2015) Histological validation of measurement of diffuse interstitial myocardial fibrosis by myocardial extravascular volume fraction from modified look-locker imaging (MOLLI) T1 mapping at 3 T. J Cardiovasc Magn Reson 17:48

Mendiola Ramírez K, Osorio Díaz JO, Maldonado Velázquez Mdel R, Faugier Fuentes E (2011) Kawasaki's disease in remission with cardiac involvement: intrasacular thrombus in a giant aneurism of both coronary arteries. Case report. Reumatol Clin 7(5):329–332

Nakahira A, Sasaki Y, Hirai H, Fukui T, Motoki M, Takahashi Y et al (2007) Rupture of aneurysmal circumflex coronary artery into the left atrium after ligation of its arteriovenous fistula. Circ J 71(12):1996–1998

Namgung J, Kim JA (2014) The prevalence of coronary anomalies in a single center of Korea: origination, course, and termination anomalies of aberrant coronary arteries detected by ECG-gated cardiac MDCT. BMC Cardiovasc Disord 14:48

Nony P, Beaune J, Champsaur G, Bozio A, Age C, Fontana J et al (1992) Anomalous origin of left coronary artery from the pulmonary artery: evolution of left ventricular function and perfusion after surgery in a 44-year-old man. Clin Cardiol 15(6):466–468

Okura N, Okuda T, Shiotani S, Kohno M, Hayakawa H, Suzuki A et al (2013) Sudden death as a late sequel of Kawasaki disease: postmortem CT demonstration of coronary artery aneurysm. Forensic Sci Int 225(1–3):85–88

Pachon R, Bravo C, Niemiera M (2015) Sudden cardiac death as a presentation of anomalous origin of the left coronary artery from pulmonary artery in a young adult. Eur Heart J Acute Cardiovasc Care 4(6):589–590

Patil S, Shirodkar S, Pinto R, Dalvi B (2008) Giant coronary artery aneurysm with a thrombus secondary to Kawasaki disease. Ann Pediatr Cardiol 1(1):59–61

Peña E, Nguyen ET, Merchant N, Dennie C (2009) ALCAPA syndrome: not just a pediatric disease. Radiographics 29(2):553–565

Quah JX, Hofmeyr L, Haqqani H, Clarke A, Rahman A, Pohlner P et al (2014) The management of the older adult patient with anomalous left coronary artery from the pulmonary artery syndrome: a presentation of two cases and review of the literature. Congenit Heart Dis 9(6):E185–E194

Raghuram AR, Krishnan R, Kumar S, Balamurugan K, Anand ND (2004) Anomalous left coronary artery from pulmonary artery (ALCAPA) in an adult. Indian J Thorac Cardiovasc Surg 20(4):213–215

Rajs J, Brodin LA, Hertzfeld I, Larsen FF (2001) Death related to coronary artery fistula after rupture of an aneurysm to the coronary sinus. Am J Forensic Med Pathol 22(1):58–61

Rana O, Swallow R, Senior R, Greaves K (2009) Detection of myocardial ischaemia caused by coronary artery-left ventricular fistulae using myocardial contrast echocardiography. Eur J Echocardiogr 10(1):175–177

Ripley DP, Gosling OE, Harries S, Spurrell PA, Bellenger NG (2014) Multimodality imaging in bland-White-Garland syndrome in an adult with a left dominant coronary artery system. Congenit Heart Dis 9(4):E110–E112

Rizk SRY, El Said G, Daniels LB, Burns JC, El Said H, Sorour KA et al (2015) Acute myocardial ischemia in adults secondary to missed Kawasaki disease in childhood. Am J Cardiol 115(4):423–427

Roberts WC, Siegel RJ, Zipes DP (1982) Origin of the right coronary artery from the left sinus of valsalva and its functional consequences: analysis of 10 necropsy patients. Am J Cardiol 49(4):863–868

Sadanandan R, Thankappan A, Jacob B, Kuriakose KM (2012) Anomalous left coronary artery from pulmonary artery (ALCAPA) repair and mitral valve replacement with bioprosthetic valve with in a 62 year old lady. Indian J Thorac Cardiovasc Surg 28(2):146–147

Safaa AM, LL D, Batra R, Essack N (2013) A rare case of adult type ALCAPA syndrome: presentation, diagnosis and management. Heart Lung Circ 22(6):444–446

Said SA (2016) Characteristics of congenital coronary artery fistulas complicated with infective endocarditis: analysis of 25 reported cases. Congenit Heart Dis 11(6):756–765

Said SAM, Schroeder-Tanka JM, Mulder BJM (2008) Female gender and the risk of rupture of congenital aneurysmal fistula in adults. Congenit Heart Dis 3(1):63–68

Sajjadieh Khajouei A, Samie-Nasab M, Behjati M, Biederman RW (2016) Cardiac computed tomography of an asymptomatic 48-year-old woman with ALCAPA syndrome. Echocardiography 33(12):1923–1925

Secinaro A, Ntsinjana H, Tann O, Schuler PK, Muthurangu V, Hughes M et al (2011) Cardiovascular magnetic resonance findings in repaired anomalous left coronary artery to pulmonary artery connection (ALCAPA). J Cardiovasc Magn Reson 13:27

Seguchi M, Nakanishi T, Nakazawa M, Doi S, Momma K, Takao A et al (1990) Myocardial perfusion after aortic implantation for anomalous origin of the left coronary artery from the pulmonary artery. Eur Heart J 11(3):213–218

Shabestari AA, Akhlaghpoor S, Tayebivaljozi R, Fattahi Masrour F (2012) Prevalence of congenital coronary artery anomalies and variants in 2697 consecutive patients using 64-detector row coronary CT angiography. Iran J Radiol 9(3):111–121

Sharples L, Hughes V, Crean A, Dyer M, Buxton M, Goldsmith K et al (2007) Cost-effectiveness of functional cardiac testing in the diagnosis and management of coronary artery disease: a randomised controlled trial. The CECaT Trial Health Technol Assess 11(49):iii–iiv, ix

Shen Q, Yao Q, Hu X (2016) Anomalous origin of the left coronary artery from the pulmonary artery in children: diagnostic use of multidetector computed tomography. Pediatr Radiol 46(10):1392–1398

Silverman NH (2015) Echocardiographic presentation of anomalous origin of the left coronary artery from the pulmonary artery. Cardiol Young 25(8):1512–1523

Sivri N, Aktoz M, Yalta K, Ozcelik F, Altun AA (2012) Retrospective study of angiographic ally determined anomalous coronary arteries in 12,844 subjects in Thrace region of Turkey. Hippokratia 16(3):246–249

Sohrabi B, Habibzadeh A, Abbasov E (2012) The incidence and pattern of coronary artery anomalies in the north-west of iran: a coronary arteriographic study. Korean Circ J 42(11):753–760

Song G, Ren W, Liu Z, Wu D (2015) Incomplete Kawasaki disease with coronary artery aneurysm and coronary sinus thrombus. Pediatr Cardiol 36(5):1097–1099

Su C-S, Tsai I-C, Lin W-W, Lin F-Y, Ting C-T, Wang K-Y (2010) Usefulness of multidetector-row computed tomography in diagnosis of anomalous origin of left coronary artery arising from the pulmonary artery. J Chin Med Assoc 73(9):492–495

Teo CHY, Paul G (2005) An uncommon cause of death in an older child part 1. Pathology 37(2):172–173

Thom H, West NEJ, Hughes V, Dyer M, Buxton M, Sharples LD et al (2014) Cost-effectiveness of initial stress cardiovascular MR, stress SPECT or stress echocardiography as a gate-keeper test, compared with upfront invasive coronary angiography in the investigation and management of patients with stable chest pain: mid-term outcomes from the CECaT randomised controlled trial. BMJ Open 4(2):e003419

Thompson WR (2015) Stress echocardiography in paediatrics: implications for the evaluation of anomalous aortic origin of the coronary arteries. Cardiol Young 25(8):1524–1530

Tobler D, Motwani M, Wald RM, Roche SL, Verocai F, Iwanochko RM et al (2014) Evaluation of a comprehensive cardiovascular magnetic resonance protocol in young adults late after the arterial switch operation for d-transposition of the great arteries. J Cardiovasc Magn Reson 16:98

Tongut A, Özyedek Z, Çerezci İ, Erentürk S, Hatemi AC (2016) Prevalence of congenital coronary artery anomalies as shown by multi-slice computed tomography coronary angiography: a single-centre study from Turkey. J Int Med Res 44(6):1492–1505

Toumpourleka M, Belitsis G, Alonso R, Rubens M, Moat N, Gatzoulis M (2015) Late presentation and surgical repair of ALCAPA. Int J Cardiol 186:207–209

Wagner A, Mahrholdt H, Holly TA, Elliott MD, Regenfus M, Parker M et al (2003) Contrast-enhanced MRI and routine single photon emission computed tomography (SPECT) perfusion imaging for detection of subendocardial myocardial infarcts: an imaging study. Lancet 361(9355):374–379

Xu H, Zhu Y, Zhu X, Tang L, Xu Y (2012) Anomalous coronary arteries: depiction at dual-source computed tomographic coronary angiography. J Thorac Cardiovasc Surg 143(6):1286–1291

Yamanaka O, Hobbs RE (1990) Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Catheter Cardiovasc Diagn 21(1):28–40

Yang Y-L, Nanda NC, Wang X-F, Xie M-X, Lu Q, He L et al (2007) Echocardiographic diagnosis of anomalous origin of the left coronary artery from the pulmonary artery. Echocardiography 24(4):405–411

Yildiz A, Okcun B, Peker T, Arslan C, Olcay A, Bulent Vatan M (2010) Prevalence of coronary artery anomalies in 12,457 adult patients who underwent coronary angiography. Clin Cardiol 33(12):E60–E64

Yuksel S, Meric M, Soylu K, Gulel O, Zengin H, Demircan S et al (2013) The primary anomalies of coronary artery origin and course: a coronary angiographic analysis of 16,573 patients. Exp Clin Cardiol 18(2):121–123