

Coronary Artery Anomalies Andrew M. Crean

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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).

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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).

		Modality of	Population	Prevalence	
First author	Country	diagnosis	size	(%)	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	2.33ª	Separate origin LAD, Cx
Yamanaka (1990)	USA	Coronary angiography	126,595	1.3ª	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

 Table 1
 Prevalence of congenital coronary anomalies in different series

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

^aDid not exclude abnormalities of high takeoff which are excluded a priori in most other series

^bDid 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

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 Kawasaki disease Vasculitic	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

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

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



caliber

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).

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.

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

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-bycase 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 freebreathing 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 mm³, 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-

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

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)

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 nonatherosclerotic 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.

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