Abnormalities of the Coronary Arteries in Children: Looking beyond the Origins[1]

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Coronary arterial abnormalities are uncommon findings in children that have profound clinical implications. Although anomalies of the coronary origins are well described, there are many other disease processes that affect the coronary arteries. Immune system–mediated diseases (eg, Kawasaki disease, polyarteritis nodosa, and other vasculiditides) can result in coronary arterial aneurysms, strictures, and abnormal tapering of the vessels. Because findings at imaging are an important component of diagnosis in these diseases, the radiologist's understanding of them is essential. Congenital anomalies may present at varying ages, and findings in hemodynamically significant anomalies, such as fistulas, are key for both diagnosis and preoperative planning. Pediatric heart surgery can result in wide-ranging postoperative imaging appearances of the coronary arteries and also predisposes patients to a multitude of complications affecting the heart and coronary arteries. In addition, although rare, accidental trauma can lead to injury of the coronary arteries, and awareness and detection of these conditions are important for diagnosis in the acute setting. Patients with coronary arterial conditions at presentation may range from being asymptomatic to having findings of myocardial infarction. Recognition of the imaging findings is essential to direct appropriate treatment.

Abbreviations: CAV = coronary allograft vasculopathy, CAVF = coronary arteriovenous fistula, KD = Kawasaki disease, LAD = left anterior descending artery, LCx = left circumflex artery, MIP = maximum intensity projection, RCA = right coronary artery, SLE = systemic lupus erythematosus, 3D = three-dimensional

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SA-CME LEARNING OBJECTIVES

After completing this journal-based SA-CME activity, participants will be able to:

- Discuss clinical and imaging features of abnormalities that may involve the pediatric coronary arteries beyond anomalous coronary arterial origins.
- Describe potential sequelae and complications of these conditions that may result in clinical symptoms, and identify imaging features of these complications.
- Identify the most appropriate imaging modality for imaging of each entity.

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Introduction

Abnormalities of the coronary arteries are uncommon yet important findings in children at imaging. Although anomalies of coronary arterial origin are relatively infrequent, with an estimated prevalence of less than 1% of the population (1), they have been extensively discussed in the literature. An understanding of the diverse disease processes (beyond anomalous origins) that may affect the coronary arteries is essential in interpretation of pediatric cardiac imaging studies. Accurate diagnosis and prompt treatment of children with these conditions can prevent adverse outcomes such as myocardial ischemia or infarction.

In this article, we discuss the approach to imaging coronary arteries in children and review the imaging features of entities other than anomalous origins that may affect the coronary arteries in children, including those that are of immune system–mediated, traumatic, congenital, and postsurgical origins.

TEACHING POINTS

- When considering immune system–mediated diseases of the coronary arteries, vascular findings should be correlated with the clinical history and laboratory data.
- Giant aneurysms in KD present a higher risk for thrombosis and infarction, and small aneurysms are more likely to regress.
- Although myocardial bridging may be an incidental finding, it is important for the radiologist to note the depth and length of the involved segment, as these factors can allow prediction of clinical significance.
- The age of the patient and severity of clinical presentation of CAVFs vary depending on the degree of coronary steal syndrome and symptoms of ischemia.
- Patients with congenital heart disease often undergo surgery that requires a coronary arterial reimplantation procedure, which is typically accomplished via a button transfer technique. In this technique, a button of aortic tissue surrounding the coronary ostium is transferred to its new location along with the coronary artery. Awareness of this procedure is important to avoid confusion with an anastomotic pseudoaneurysm.

Imaging Approach

Conventional angiography is recognized as the standard for diagnosis of many conditions affecting the coronary arteries in children. Although conventional angiography allows for excellent evaluation of the vessel lumen, it is limited because it does not clearly show the relationship between the coronary arteries and adjacent structures. In addition, it has the potential to impart high doses of ionizing radiation. Other disadvantages include procedure-related complications such as pseudoaneurysm, dissection, and stroke, as well as the need for specialized equipment and personnel.

Echocardiography is excellent for visualizing cardiac chambers. However, smaller structures such as the coronary arteries may not be as well seen. Adequate visualization of the coronary arteries at echocardiography may be hindered in larger children. As echocardiography does not use ionizing radiation, it is useful in patients who require repeated screening. It may also be useful in settings in which evaluation of cardiac function or wall motion is required.

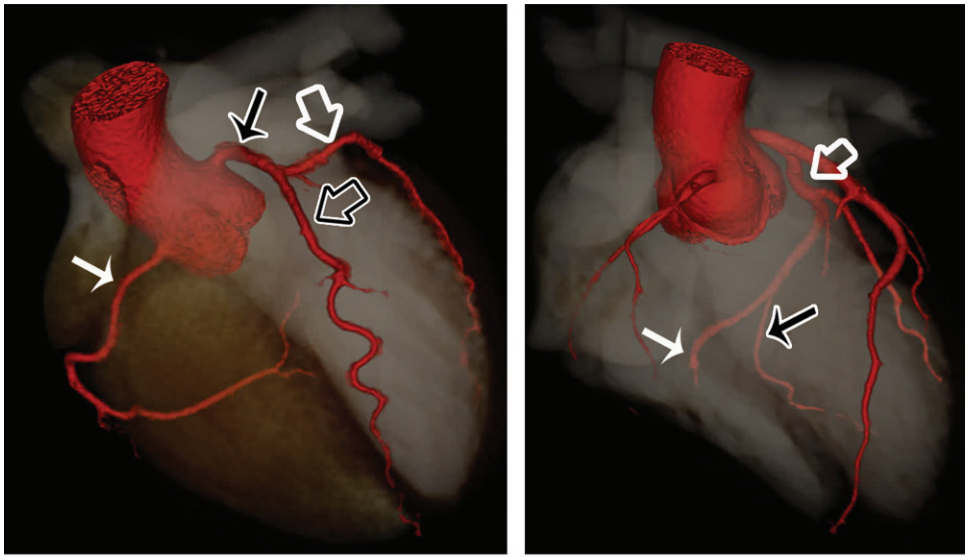
Although magnetic resonance (MR) angiography is not generally used for dedicated imaging of the coronary arteries, coronary arterial origins can typically be seen, particularly when an intravenous gadolinium-based contrast agent is used. Although blood pool agents such as gadofosveset were previously useful for vascular MR imaging examinations, their current limited availability from manufacturers has prompted the exploration of other agents for this purpose, including small superparamagnetic iron oxide particles such as ferumoxytol. Smaller, more distal arteries may remain difficult to visualize even at angiographic examination, especially in pediatric patients.

In the era of multidetector computed tomography (CT), coronary CT angiography is rapidly becoming the preferred technique for anatomic evaluation of the coronary arteries in children. In addition to the excellent spatial resolution and less-invasive nature of CT, this modality demonstrates the relationship of the coronary arteries to adjacent structures and is fast, often negating the need for sedation. Data have shown excellent correlation between findings at CT angiography and conventional angiography (2,3). Despite there being less ionizing radiation than from conventional angiography, consideration must be given to techniques to keep the CT dose as low as possible in children, including reduced kilovoltage, higher pitch, and the routine use of iterative reconstruction. An understanding of coronary arterial anatomy as seen at CT angiography is essential for interpretation of these examinations. A review of the relevant anatomy is presented in Figure 1.

Immune System–mediated Conditions

Immune system–mediated pathologic conditions of the coronary arteries are varied in clinical presentation, and imaging findings may present similar appearances in multiple entities. Clinical findings are often nonspecific and may make diagnosis challenging. When considering immune system–mediated diseases of the coronary arteries, vascular findings should be correlated with the clinical history and laboratory data. The most common vasculitides affecting coronary arteries in children are Kawasaki disease (KD) and polyarteritis nodosa. Rarely, other immune system–mediated processes, including Takayasu arteritis and systemic lupus erythematosus (SLE), can affect the coronary arteries. In the setting of heart transplant, coronary allograft vasculopathy (CAV) should be considered.

Kawasaki Disease.—KD is the second most common childhood vasculitis after Henoch-Schönlein purpura and the most common vasculitis affecting the coronary arteries (4). The peak age at onset is 2–3 years, and it is more common in boys, with a male-to-female ratio of from 1.5:1 to 2:1. In the acute phase, patients present with a vague clinical syndrome that includes fever, conjunctivitis, rash, and cervical lymphadenopathy. In the subacute phase of KD, the fever resolves and patients are at their highest risk for development of coronary arterial aneurysms and sudden cardiac death related to acute thrombosis and infarction (5,6). The convalescent phase begins at the end



a. b.

Figure 1. **(a)** Three-dimensional (3D) volume-rendered image from coronary CT angiography in a 17-year-old adolescent girl with right-dominant anatomy. The left main coronary artery (solid black arrow) arises from the left sinus of Valsalva and divides into the left anterior descending artery (LAD) (open black arrow) and left circumflex artery (LCx) (open white arrow). Diagonal branches arise from the LAD, supplying the anterior and anteroseptal myocardium, apex, and anterior two-thirds of the interventricular septum. Obtuse marginal branches arise from the LCx, supplying the anterolateral and inferolateral left ventricular wall (these smaller branches are not well seen on this 3D volume-rendered image). The right coronary artery (RCA) (solid white arrow) arises from the right sinus of Valsalva and courses in the interventricular groove, supplying the right ventricle. Coronary arterial dominance is defined by which coronary artery gives rise to the vessels that supply the inferior aspect of the heart. In this right-dominant heart, the RCA gives rise to the posterior descending artery and posterolateral ventricular branch (PLVB) and supplies the posterior one-third of the interventricular septum and inferior wall of the left ventricle. **(b)** Three-dimensional volume-rendered image from coronary CT angiography in a 19-year-old man with left-dominant anatomy. Left- or codominant anatomy occurs in 15%–20% of the population. The LCx (open white arrow) gives rise to the posterior descending artery (black arrow) and PLVB (solid white arrow), supplying the inferior wall of the left ventricle and posterior one-third of the interventricular septum. In codominant hearts, the posterior descending artery is supplied by the LCx, and the PLVB is supplied by the RCA.

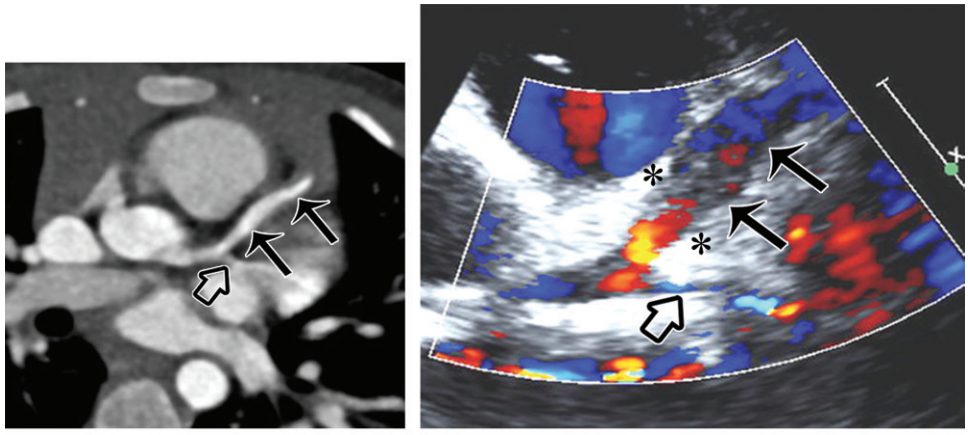
of the subacute phase, typically 4–8 weeks after onset of the illness.

Cardiovascular complications are the leading cause of morbidity and mortality in KD (7). Echocardiography is the primary imaging modality used to screen for coronary arterial aneurysms in patients with KD. Echocardiography is preferred for screening because of its lack of ionizing radiation and typically allows visualization of the proximal coronary arteries, where aneurysms are most commonly found.

Features of KD can also be seen at MR angiography and CT angiography. These modalities may be more useful in evaluating the distal coronary arteries, especially in older or larger children with more limited echocardiographic windows (8–10). Use of MR angiography or CT angiography in these settings may obviate the use of conventional coronary angiography, which is more invasive (10). Given its higher spatial resolution and shorter examination time, CT angiography is the preferred modality for KD diagnosis (11,12). A potential benefit of MR angiography is that it can enable evaluation for associated findings such as myocarditis and pericarditis and assessment of myocardial function.

Imaging findings of KD include coronary arterial ectasia and aneurysms, lack of normal tapering, and increased perivascular echogenicity (13) (Figs 2, 3). Coronary arterial aneurysms in KD are more likely to develop in the proximal coronary arterial systems, with the proximal LAD and proximal RCA being the most common sites. Distal aneurysms are unusual in the absence of proximal disease (7). Aneurysms are classified as small (<5-mm internal diameter), medium (5–8 mm), and giant (>8 mm). Giant aneurysms in KD present a higher risk for thrombosis and infarction, and small aneurysms are more likely to regress (14,15). The calcifications that can be seen within the periphery of aneurysms in KD are seen to better advantage at CT angiography and may be easily missed at MR angiography.

Other cardiac findings in KD include myocarditis, which histologic evidence suggests is a nearly universal finding, and pericarditis (7). Mitral or



a. b.

Figure 2. Fusiform aneurysmal dilatation in a 4-year-old boy with KD. Curved planar reformation **(a)** from coronary CT angiography along the LAD and color Doppler echocardiogram **(b)** show long-segment fusiform aneurysmal dilatation (solid black arrows) of the LAD, measuring 5 mm at its greatest internal diameter. Perivascular echogenicity (∗ in **b**) and the proximal LCx (open arrow) are also demonstrated.

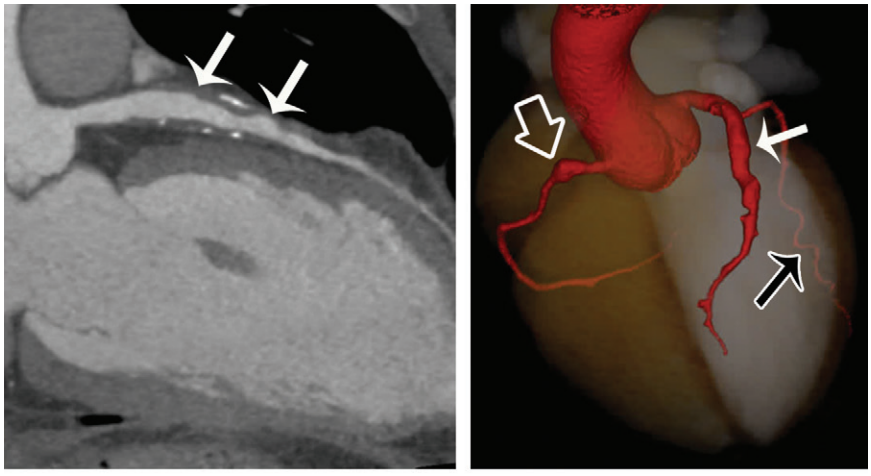


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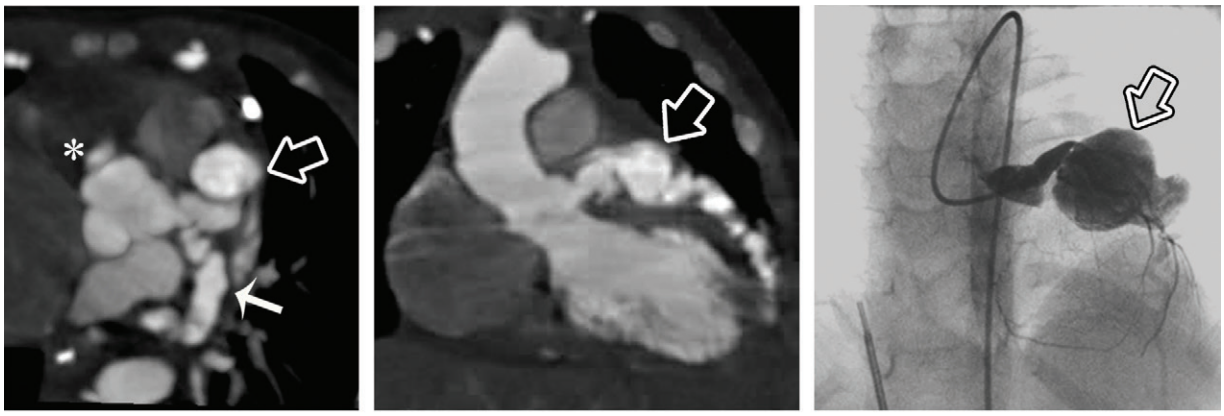
Figure 3. Fusiform aneurysmal dilatation in a 17-year-old adolescent girl with a remote history of KD. **(a)** Curved planar reformation from coronary CT angiography along the LAD shows a giant (14-mm internal diameter) coronary arterial aneurysm with extensive long-segment fusiform aneurysmal dilatation of the LAD (arrows) with eccentric mural thrombus, containing calcifications. **(b)** Three-dimensional volume-rendered image shows extensive fusiform dilatation (solid white arrow) of the LAD and a more focal fusiform aneurysm (open white arrow) of the proximal RCA. The LCx (black arrow) courses behind the left ventricle on this view.

aortic valvular regurgitation may occur secondary to associated ischemia or valvulitis or to transient papillary muscle dysfunction involving the mitral valve (7,16). Extracardiac manifestations may include colitis and gallbladder hydrops (17). Systemic aneurysms may also be seen in KD but only in patients with coronary aneurysms (14).

Polyarteritis Nodosa.—Polyarteritis nodosa is a rare childhood necrotizing inflammatory vasculitis of medium or small vessels. The mean age at onset in children is approximately 9 years, with no sex predilection (18,19). Presenting symptoms are nonspecific and include fever, myalgia, arthralgia, and skin lesions (18). The most commonly involved organ systems are the skin, renal, and gastrointestinal systems; cardiac involvement is less common (13,18). At laboratory analysis, patients may have positive hepatitis B virus (HBV) surface antigen, although HBV-associated polyarteritis nodosa is less common in children (13).

Involvement of medium-sized visceral arteries is characteristic in polyarteritis nodosa, primarily involving the renal, hepatic, and mesenteric arterial vasculature. Aneurysms of the visceral arterial vasculature at conventional angiography are generally accepted as diagnostic (13). Other findings include arterial beading and caliber variations, arterial tapering, stenosis, occlusion, and pruning of the peripheral vasculature (20).



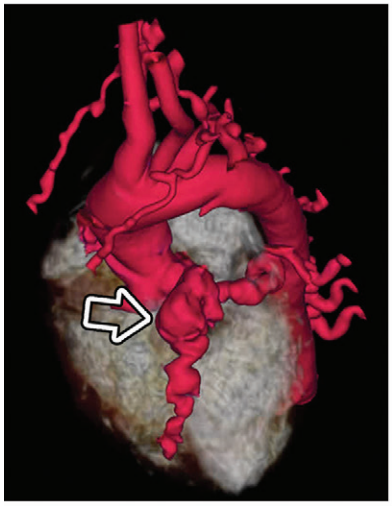


Figure 4. Saccular and fusiform aneurysms in a 10-year-old boy with polyarteritis nodosa. **(a)** Axial maximum intensity projection (MIP) image from coronary CT angiography demonstrates complete thrombosis (*) of the RCA and saccular and fusiform aneurysms of the LAD (open arrow) and LCx (solid arrow). **(b)** Curved planar reformation along the LAD shows the extent of the aneurysms (arrow), which extend to the left ventricular apex. **(c)** Correlative conventional angiographic image demonstrates the saccular nature of the largest, proximal LAD aneurysm (arrow). **(d)** Three-dimensional volume-rendered image redemonstrates the coronary arterial findings in the LAD (arrow), as well as multiple areas of dilatation in branch arteries.

Cardiac involvement is less common and typically occurs in the setting of visceral involvement. Although only 5%–20% of patients have symptoms of cardiac involvement (21), autopsies have shown evidence of coronary arteritis at pathologic analysis in nearly half of patients with polyarteritis nodosa (22). Cardiac manifestations include coronary arteritis, pericarditis, and, rarely, myocarditis (21). Congestive heart failure can occur as a result of ischemia secondary to coronary arterial vasculitis, hypertension, or both (21,23). Heart failure has also been shown to occur in the setting of normal coronary arterial imaging (18). When heart failure is present, typical imaging findings in the coronary arteries mimic those in the abdomen and include vessel caliber variation, thrombosis, and aneurysmal dilatation, often better seen at conventional or CT angiography, given that the findings are often present in the mid- and distal arteries. The characteristic "beads on a string" sign often seen in polyarteritis nodosa in the mesenteric vessels may be present at coronary angiography as well (21) (Fig 4).

Takayasu Arteritis.—Takayasu arteritis is a large-vessel vasculitis that predominantly affects the aorta and its main branches. Although it typically affects young women, children may also be affected, although rarely. Childhood Takayasu arteritis typically manifests between 10 and 20 years of age, most commonly with hypertension (24).

Tann et al (25) estimated that 10%–20% of adult patients with Takayasu arteritis have coronary arterial involvement, and multiple case reports exist of coronary arterial involvement in children (26–28). In adults, the most common sites of involvement are the coronary arterial ostia and proximal segments (29). Although there are no large studies of the imaging findings of coronary arterial involvement in Takayasu arteritis in children, reported cases have described perivascular hyperechogenicity at echocardiography (28) and ostial and proximal coronary artery stenosis, which can be severe (26–28).

Systemic Lupus Erythematosus.—Pediatric SLE accounts for 10%–20% of patients with SLE, with an incidence of approximately 3.3 to 24 cases per 100 000 children (30). Pediatric patients typically present between 12 and 16 years of age. The female preponderance that is present in the adult SLE population is much less pronounced in children. SLE manifesting in childhood tends to be more severe, with higher rates of solid-organ than mucocutaneous or hematologic involvement and a more aggressive clinical course (31). Common presenting features of lupus in children include

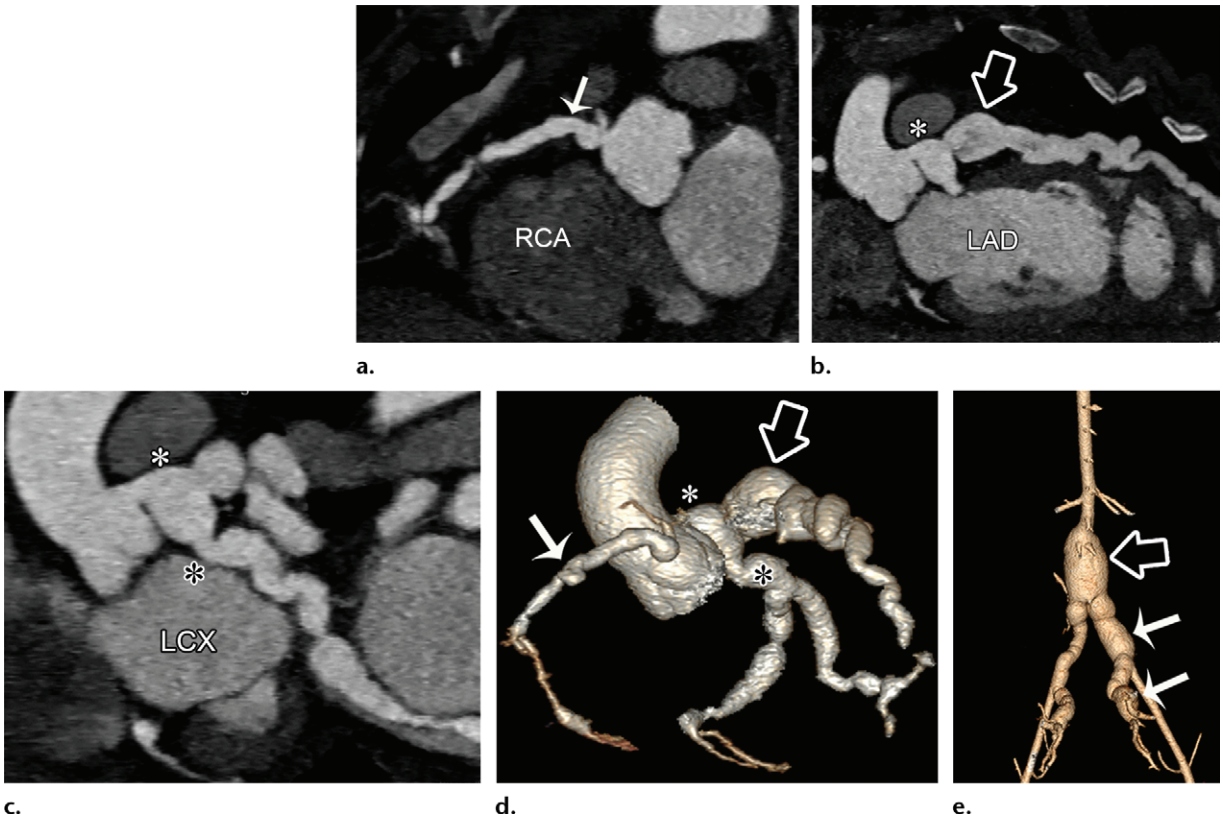


Figure 5. Multifocal aneurysmal dilatation in a 9-year-old girl with SLE. **(a–d)** Curved planar reformatted images **(a–c)** and 3D volume-rendered image **(d)** from coronary CT angiography demonstrate multifocal aneurysmal dilatation of the RCA (solid arrow in **a** and **d**), left main coronary artery (white ∗ in **b–d**), LAD (open arrow in **b and d**), and LCx (black ∗ in **c** and **d**). **(e)** Three-dimensional volume-rendered image demonstrates aneurysmal dilatation of the infrarenal aorta (open arrow) and internal iliac arteries (solid arrows).

nephropathy, neuropsychiatric disease, arthritis, fever, malar rash, and leukopenia (30).

Although the most common cardiac manifestation of SLE is pericarditis, other structures may be affected, including the coronary arteries, myocardium, and cardiac valves (32). In adult patients, coronary arterial involvement in SLE typically consists of premature atherosclerosis, whereas coronary arterial vasculitis is more common in children (32,33). Rates of coronary arteritis in childhood lupus may range from 4% to 10% (32,33).

Coronary arteritis in SLE appears similar in both children and adults and is characterized by aneurysmal dilatation, both focal and diffuse, with or without areas of stenosis (33–35) (Fig 5). These findings have been more commonly demonstrated at echocardiography (34) and conventional angiography (35), although similar findings are visible at CT (36). Following treatment, the coronary arterial lesions may improve or resolve completely (34). In severe cases, coronary arterial occlusion leading to ischemia or infarction can occur (32,35,37). This is typically secondary to vasculitis in children (32,33).

Coronary Allograft Vasculopathy.—CAV is a form of coronary arterial condition in heart transplant recipients characterized by concentric fibrous intimal hyperplasia (38). Although diagnosis can be challenging, CAV is an important cause of graft failure in transplant recipients, as graft survival is approximately 50% at 5 years after diagnosis (39).

Presenting features of CAV are variable and can include multiple episodes of rejection, arrhythmia, or conduction abnormalities, as well as recurrent chest or abdominal pain. Owing to variations in reinnervation of the allograft in recipients, patients with ischemia or infarction in the setting of CAV may present with atypical chest pain or no chest pain (39). Patients may also demonstrate systolic or diastolic dysfunction in the absence of rejection at presentation, and the first symptom of CAV may be sudden death (40).

Pathologic changes include concentric intimal hyperplasia, vasculitic changes, and thrombus formation. These findings may affect not only the coronary arterial vasculature but also the coronary veins and can be seen in vessels of all calibers, from large epicardial vessels to intramyocardial microvasculature. These features help distinguish this entity from atherosclerosis (38).

The pathologic findings of CAV in children are reflected in its imaging appearances, classically including diffuse irregularity and nar-

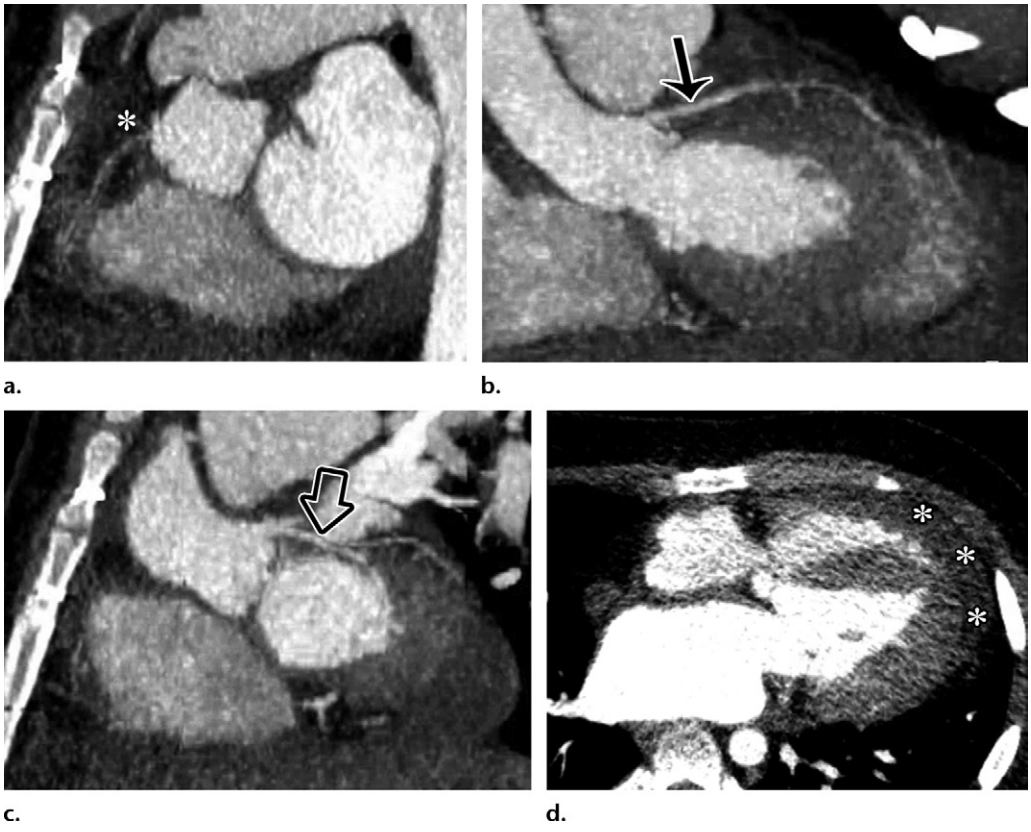


Figure 6. CAV in a 16-year-old adolescent boy with a history of orthotopic heart transplant 4 years previously for dilated cardiomyopathy, now with worsening diastolic dysfunction. **(a–c)** Curved planar reformatted images from coronary CT angiography demonstrate a diffusely diminutive RCA (* in **a**), LAD (arrow in **b**), and LCx (arrow in **c**) with diffuse mural thickening and luminal narrowing. **(d)** Axial coronary CT angiographic image shows pericardial effusion (*). Although pericardial effusion has not been described in the setting of CAV, it may occur for many reasons in the posttransplant patient, including as a result of rejection (43) or secondary to immunosuppressant medications (44).

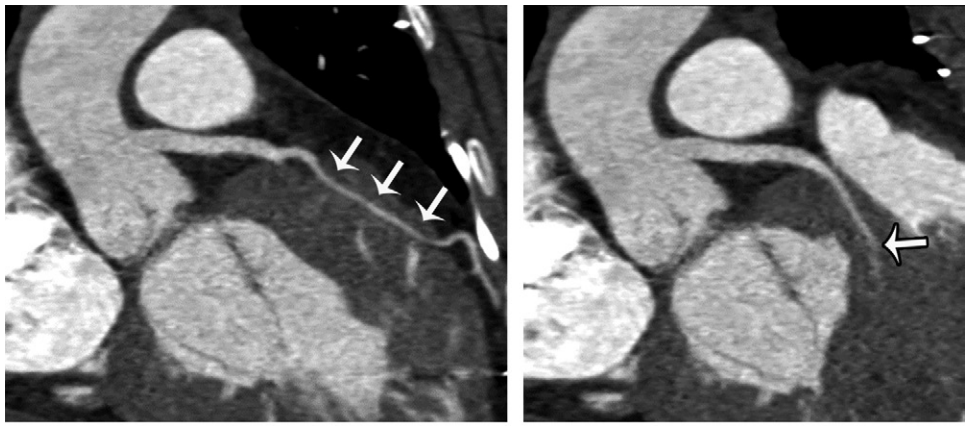
rowing of the epicardial vessels or pruning of the distal vessel caliber, typically without focal stenoses (39). The current standard for diagnosis is conventional angiography, which generally has high specificity with moderate sensitivity (41). It may be difficult to detect subtle changes, and angiography remains an operator-dependent technique that is technically challenging in small patients. Findings similar to those seen at conventional angiography can be identified at CT angiography (42) (Fig 6) and MR angiography (45). Although data are limited, MR angiography may also show late gadolinium enhancement in a subendocardial or transmural pattern typical of infarct or enhancement of the coronary artery vessel wall (45).

Many additional diagnostic parameters for CAV have been explored in echocardiography, including decreased ejection fraction (46), development of restrictive physiology and wall motion abnormalities (47,48), and reduced peak systolic velocities (49). Although the role of echocardiography is evolving, coronary angiography remains the standard for diagnosis.

Congenital Conditions

Congenital anomalies of the coronary arteries are uncommon, with an estimated prevalence of 0.2%–1.2% (50). The age of the patient and severity of clinical presentation depend on the degree of hemodynamic impact, as not all congenital coronary arterial anomalies are symptomatic. Those that are not hemodynamically significant may be incidentally detected. Although anomalies of coronary arterial origin fall within this group, they are not discussed in this article because of the abundance of previously published literature on this topic. Shriki et al (51) offer a comprehensive review of anomalies of coronary arterial origins.

Myocardial Bridge.—Myocardial bridge is the most common coronary arterial variant. It occurs when a coronary arterial segment is located partially or completely within the myocardium. The most common location for myocardial bridge is within the midsegment LAD, followed by the distal LAD (52). In most patients, myocardial bridge is considered to be a normal variant (52,53). However, some authors have suggested



a. b.

Figure 7. Myocardial bridging in a 16-year-old adolescent girl with hypertrophic cardiomyopathy. **(a)** Curved planar reformation from coronary CT angiography shows a long segment of deep myocardial bridging (arrows) of the LAD. **(b)** Image from the same study in a slightly different plane shows a septal perforating artery (arrow) that is distinct from the bridging and terminates in the myocardium, a normal finding.

that it may not be an entirely benign entity (52), as there may be an association with myocardial ischemia, infarction, and sudden death (54). The prevalence of myocardial bridging is much higher in patients with hypertrophic cardiomyopathy (about 30%) than in the general population (1%–3%) (55), and myocardial bridging is a risk factor for myocardial ischemia and sudden death in these patients (56).

The likelihood that a myocardial bridge will become symptomatic increases with increased length and depth of the bridged segment, which in turn influence the degree of systolic compression (57,58). When systolic compression is severe, it may impair diastolic relaxation, resulting in decreased diastolic flow distally (52,58). Increased sympathetic tone or tachycardia resulting in decreased duration of diastole may also increase the likelihood of ischemia (58). Although the bridged portion of the artery is protected from atherosclerosis, accelerated atherosclerosis can be seen proximal to the bridge later in life (59).

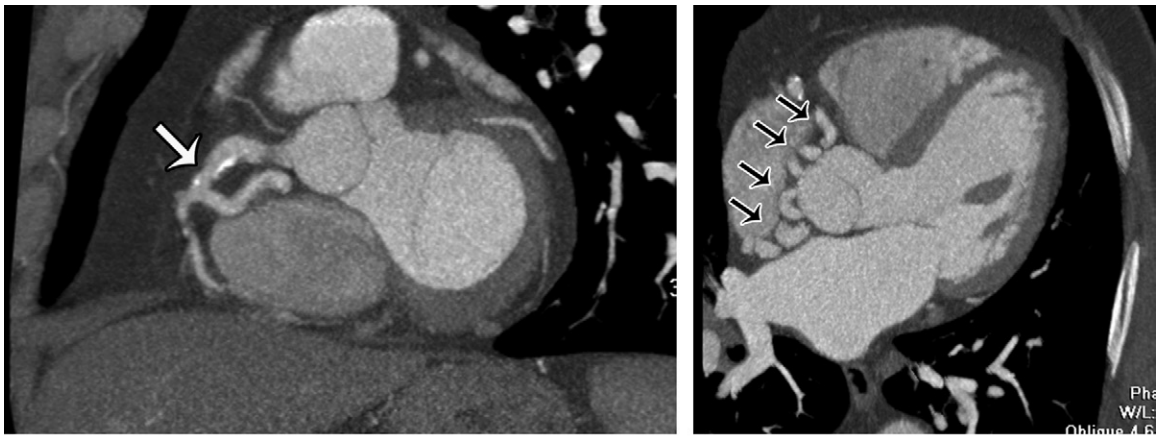
CT angiography can delineate the anatomic location, length, and depth of the myocardial bridge and is the preferred imaging modality. It is more sensitive than conventional angiography, which only depicts bridges that are deep or demonstrate significant systolic compression (53,58). Myocardial bridging should be distinguished from cases of septal perforating arteries, which terminate in the myocardium. Although myocardial bridging may be an incidental finding, it is important for the radiologist to note the depth and length of the involved segment, as these factors can allow prediction of clinical significance (Fig 7).

Fistula.—Coronary arteriovenous fistulas (CAVFs) are abnormalities of coronary arterial termination. They are a rare anomaly, with an estimated prevalence of 0.002% in the general population, but comprise 48.7% of all congenital coronary anomalies and are the most common hemodynamically significant congenital coronary lesion (60). Rarely, a fistula may be acquired, secondary to iatrogenic, traumatic, or disease-related processes. However, these instances are more commonly seen in the adult population.

Low-pressure structures are the most common sites of drainage of CAVFs (61). CAVFs originate from the RCA in approximately 50% of patients, from the left coronary artery in approximately 42% of patients, and from both the RCA and left coronary artery in 5% of patients. The most common drainage sites for a CAVF are the right ventricle (41%), right atrium (26%), and pulmonary artery (17%). Less common drainage sites include the coronary sinus, left atrium, left ventricle, and superior vena cava (62).

Drainage of a CAVF into a right-sided structure results in a left-to-right shunt, which occurs in up to 90% of patients (63). Although the majority of adult patients are asymptomatic and may have fistulas that are incidentally detected, a larger percentage of pediatric patients are symptomatic (61). The age of the patient and severity of clinical presentation of CAVFs vary depending on the degree of coronary steal syndrome and symptoms of ischemia. Presenting symptoms range from a murmur or arrhythmia to symptoms of congestive heart failure, stroke, or myocardial ischemia or infarction (61).

Diagnosis of CAVF at imaging may be accomplished via conventional, CT, or MR angiography. Echocardiography can be used to identify shunting via a bubble study, in which agitated saline is injected intravenously and in the presence of shunt-



a. b.

Figure 8. CAVF terminating in the right atrium in a 27-year-old woman. **(a)** Curved planar reformatted image from coronary CT angiography demonstrates that the fistula arises as a branch from the proximal RCA. Calcifications (arrow) within the RCA were presumed to be related to a high-flow high-pressure state secondary to the fistula. **(b)** Axial image from the same study shows that multiple tortuous dilated vessels (arrows) are present.

ing will appear more rapidly in the left ventricle than in patients without shunts. However, echocardiography is limited in its ability to define anatomy. Cardiac MR angiography provides the potential to assess blood flow volume, cardiac function, and shunt fraction (64). However, it requires longer examination times and has poorer spatial resolution than does CT angiography (63). Shorter examination time, noninvasiveness, and high temporal and spatial resolution make CT angiography a good alternative to coronary angiography (64). It can be used to identify the origin and course of the fistula, assess its complexity, and perform preoperative planning. Three-dimensional CT reconstructions, which allow easy visualization of the relationship of the fistula to adjacent structures, often prove helpful in the preoperative assessment.

Abnormally draining coronary arteries often appear enlarged and tortuous (Fig 8) (51). Arterial phase contrast opacification in the receiving chamber can allow confirmation of the fistula entry site. Although coronary angiography can reliably enable detection of the proximal portions of CAVFs, the distal drainage site may not be well demonstrated, owing to contrast material dilution when draining into low-pressure structures (65). The relationship of the fistula to adjacent anatomic structures is also better defined at CT angiography.

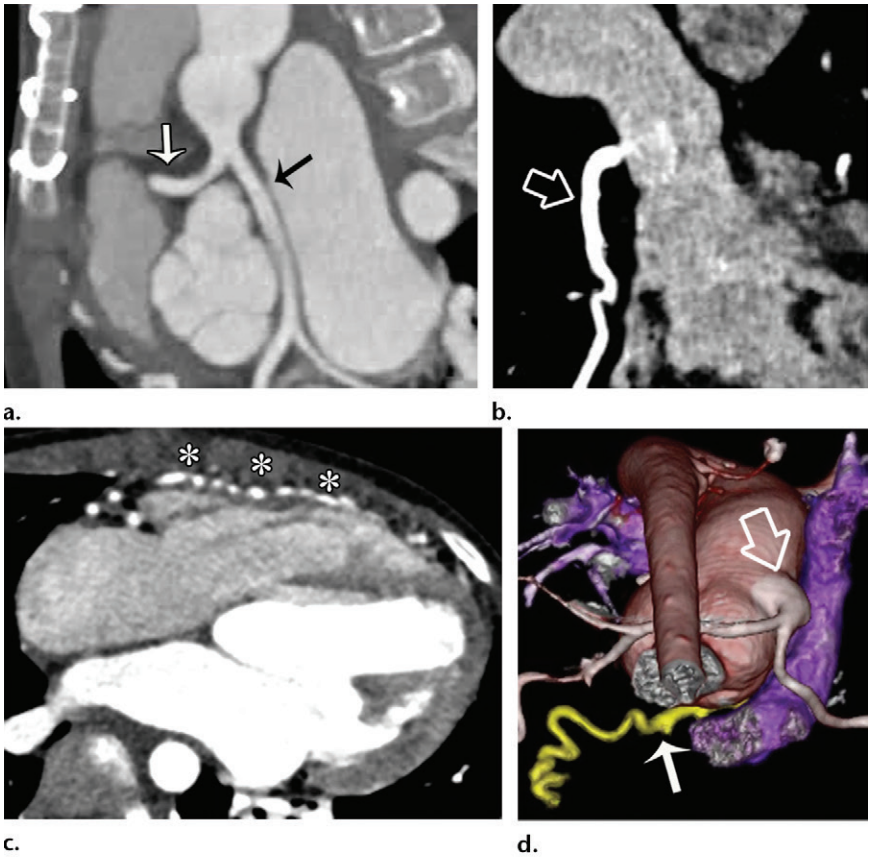
It should be noted that coronary arteries that terminate in the systemic circulation typically do not result in this pattern, as there is usually no pressure difference between the coronary artery and the systemic artery in which it terminates. These fistulas also tend to have a very small or insignificant shunt fraction (51).

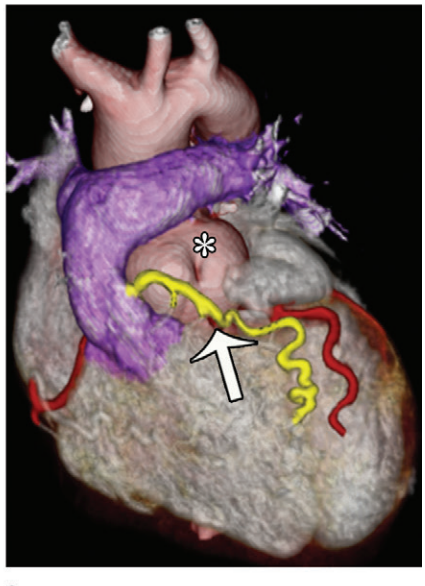
Duplication.—Coronary arterial duplication is a rare anomaly that most commonly affects the LAD (51). Typically a dual LAD consists of a short LAD that ends proximally in the interventricular groove and a long LAD that enters the interventricular groove distally and follows the expected course of the mid and distal LAD segments. The long LAD may originate as a proximal branch of the LAD or originate anomalously, often as a branch from the RCA (2,66). Duplicated coronary arteries are important to recognize at imaging as they may present implications for surgical planning. They can easily be overlooked at surgery, which could result in inadvertent transection or incomplete correction of other congenital anomalies (Fig 9). Duplicated coronary arteries are best identified at CT angiography and may be missed at conventional angiography because of failure to cannulate the second LAD. Care should be taken by the radiologist to follow the LAD along its entire course to ensure that only a single one exists.

Postsurgical Conditions

Children may undergo coronary arterial surgery for congenital cardiac abnormalities, including anomalous coronary arterial origins and complex congenital heart disease. Knowledge of the patient's history and of common surgical techniques is essential for interpretation of postoperative images. Patients with congenital heart disease often undergo surgery that requires a coronary arterial reimplantation procedure, which is typically accomplished via a button transfer technique. In this technique, a button of aortic tissue surrounding the coronary ostium is transferred to its new location along with the coronary artery. Awareness of this procedure is important to avoid confusion with an anastomotic pseudoaneurysm (Fig 10c). In addition, surgical intervention performed on

Figure 9. Double-outlet right ventricle with dextro-malpositioned great vessels in a 7-year-old girl after repair with an arterial switch procedure. **(a, b)** MIP curved planar reformatted images from coronary CT angiography demonstrate findings of arterial switch with reimplantation of a single coronary artery on the posterior sinus of the neoaorta. **(a)** The single coronary artery bifurcates into the right (white arrow) and left (black arrow) main coronary arteries. **(b)** A duplicated second LAD (arrow) that was overlooked at the initial surgery and arises from the neo–pulmonary artery is faintly opacified with arterial phase contrast agent, likely owing to a component of steal phenomenon, which occurs due to preferential flow of blood into the lower-pressure pulmonary arterial system, thus stealing flow from the myocardium. However, at surgery, a tight stenosis was found at the orifice of this duplicated coronary artery, likely limiting the effects of steal. **(c)** Axial image from the same study demonstrates multiple collateral vessels (*). **(d, e)** Findings are redemonstrated on 3D reformatted images. The duplicated LAD (solid arrow) arising from the neo–pulmonary artery is highlighted in yellow. The single main coronary artery (open arrow in **d**) arising from the neoaorta (* in **e**) after the switch procedure is also redemonstrated.

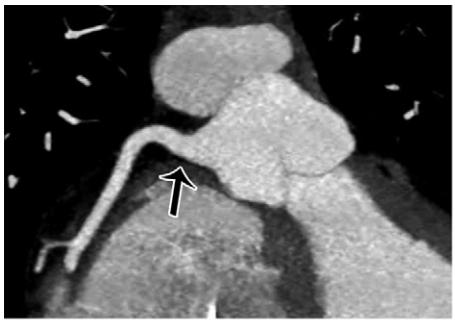




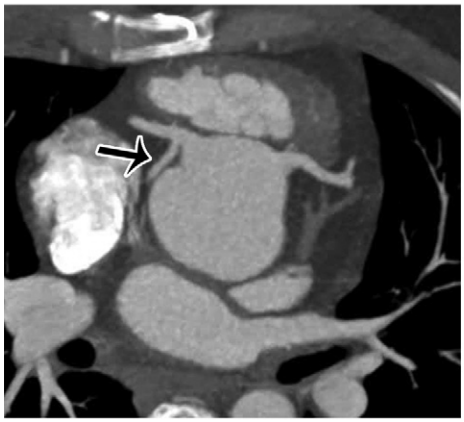
the heart in children may result in iatrogenic injury to the coronary arteries. Iatrogenic coronary arterial injury or transection should be suspected in postoperative patients who present with symptoms of cardiac ischemia (Fig 11).

Traumatic Conditions

Cardiac injury in children in the setting of trauma is uncommon and typically occurs in the setting of extensive multisystem trauma (67). Reported rates of cardiac injury in blunt trauma range from 4.6% to 7.7% (68,69). Cardiac injury in penetrating trauma is exceedingly rare. The most common traumatic cardiac injury in children is cardiac contusion, typically secondary to blunt cardiac injury. Although traumatic cardiac injury, including contusion and coronary arterial injury, may be clinically



a.



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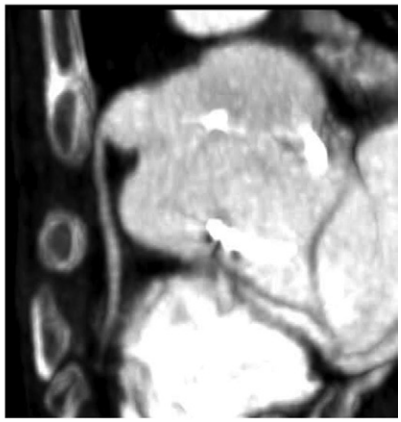


c.

Figure 10. Postoperative imaging appearances in three patients with a history of coronary arterial reimplantation. **(a)** A 22-year-old man with a history of dextro-transposition of the great vessels (D-TGA) after a coronary arterial switch procedure. Coronal oblique MIP image from coronary CT angiography demonstrates mild broadening (arrow) of the origin of the RCA at the level of reimplantation secondary to the button reimplantation technique. **(b)** A 17-year-old male adolescent with a history of D-TGA after an arterial switch procedure. MIP image from coronary CT angiography demonstrates reimplantation of both coronary arteries onto a single cusp. Note the retroaortic circumflex artery (arrow) arising from the RCA. **(c)** A 21-year-old man with a history of Loeys-Dietz syndrome after aortic root and valve replacements with button reimplantation of the coronary arteries. Oblique MIP image from coronary CT angiography demonstrates apparent aneurysmal dilatation of the button of aortic tissue at the level of the anastomosis, which can be a normal postoperative appearance.

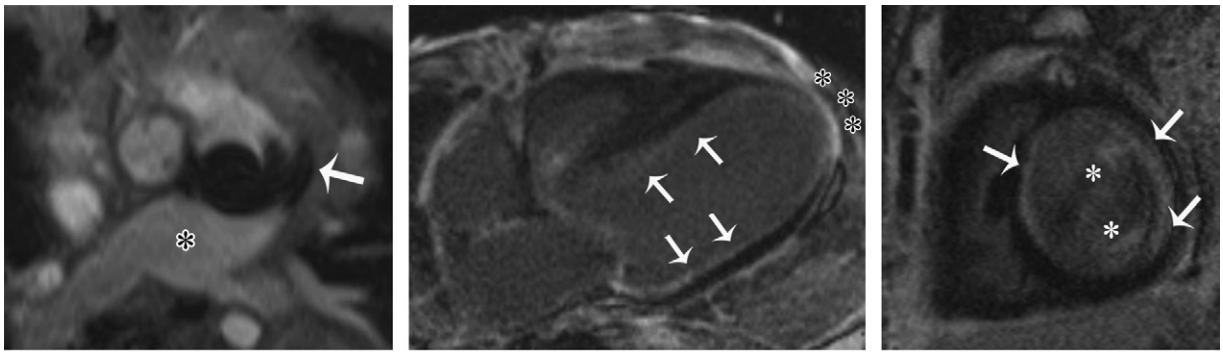
silent, electrocardiographic (ECG) abnormalities, cardiac enzyme elevation, or wall motion abnormalities at ECG may indicate the presence of coronary arterial injury in children with trauma (68,70–72).

Coronary arterial dissection in the setting of blunt trauma has been described in both adult and pediatric patients, with four case reports since 1998 of coronary arterial dissection in children (73). Some 76% of traumatic dissections involve the LAD (72). Imaging features of traumatic coronary arterial dissection are similar to those of dissection in other arteries, with luminal irregularity and narrowing of the involved artery. The small size of the coronary arteries can make visualization of these findings difficult at MR or CT angiography (Fig 12). More often, the injury results in coronary arterial occlusion and myocardial ischemia, which lead to wall motion abnormalities or valvular regurgitation. These findings are often the initial clue to the diagnosis (70–72).

CAVF, laceration, or pseudoaneurysm are more commonly features of penetrating trauma, although they can also be seen in the setting of blunt trauma (74,75). Posttraumatic fistulas most commonly arise from the LAD and terminate in low-pressure chambers such as the atria (69,74). Penetrating coronary arterial injury may also result in hemopericardium, tamponade, and rapid clinical decompensation (69). Traumatic fistula can be confirmed at Doppler echocardiography or angiography, which demonstrates a direct connection between the coronary artery and the involved cardiac chamber. Traumatic CAVF has an excellent prognosis after prompt repair, which can be accomplished surgically or percutaneously (75).

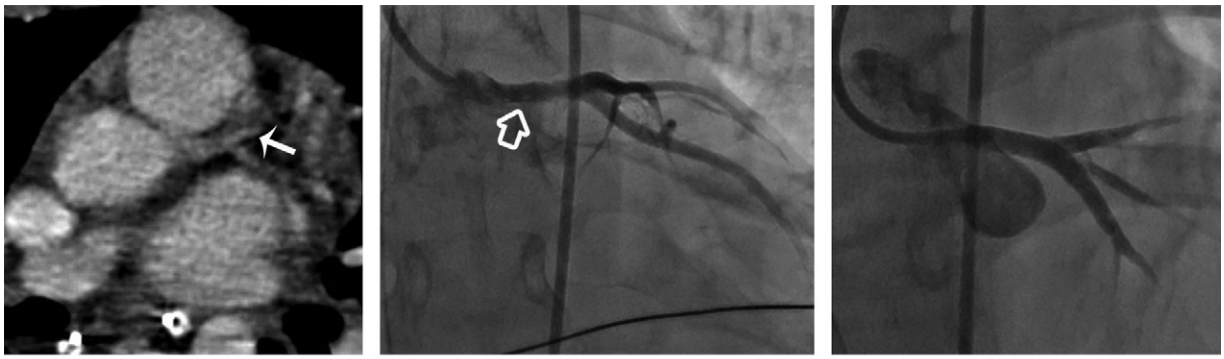
Conclusion

Pediatric coronary arteries may be affected by immune system–mediated disease, congenital abnormalities, trauma, or surgery. Immune



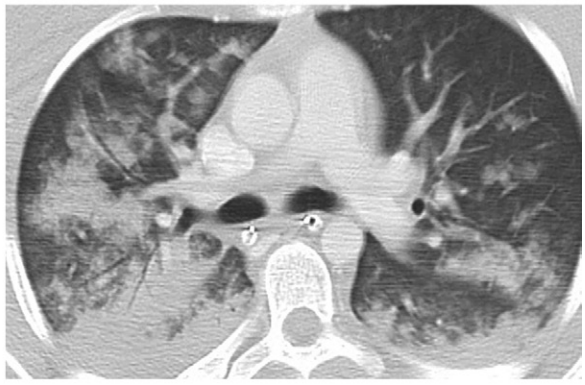
a. b. c.

Figure 11. Iatrogenic injury in a 7-year-old boy with tetralogy of Fallot who demonstrated cardiogenic shock with mitral regurgitation at presentation. The patient had undergone open surgical pulmonary valve replacement approximately 2 months previously. The surgery was complicated by cardiac arrest at termination of the procedure, secondary to transection of a single congenitally anomalous coronary artery. **(a)** Axial balanced steady-state free precession image at the level of the main pulmonary artery shows susceptibility artifact (arrow) secondary to changes of pulmonary valve replacement, with dilatation (*) of the main pulmonary artery. **(b)** Four-chamber delayed postcontrast image shows diffuse subendocardial delayed contrast enhancement (arrows) of the anterolateral wall, inferoseptal wall, and apex, with marked apical thinning (*). **(c)** Two-chamber short-axis delayed postcontrast image redemonstrates diffuse subendocardial delayed contrast enhancement (arrows), including delayed enhancement (*) of the left ventricular papillary muscles, with relative sparing of the inferior wall, consistent with myocardial infarction following iatrogenic transection.



a. c. d.

Figure 12. Traumatic cardiac injury in a 14-year-old boy. Blunt trauma to the chest from another player's knee while playing basketball resulted in ventricular arrhythmia and pulselessness, with an elevated troponin level of greater than 62 ng/mL. **(a)** Non–electrocardiographically-gated chest CT image shows possible narrowing (arrow) of the left main coronary artery and proximal LAD. **(b)** Axial CT image at the level of the heart shows pulmonary edema. **(c)** Conventional angiogram shows mural irregularity and narrowing of the left main coronary artery extending into the LAD (arrow), consistent with dissection. **(d)** Image after angioplasty and stent placement shows a widely patent left main coronary artery and LAD.



b.

system–mediated diseases may share similar imaging findings; correlation with the clinical presentation and laboratory data is essential. Patient presentation and imaging findings may vary depending on the degree of hemodynamic significance of the lesion, particularly in congenital conditions. Surgical procedures can result in iatrogenic injury with resultant ischemia. Surgery in the setting of congenital heart disease can have a characteristic imaging appearance and must be distinguished from abnormality. Coronary arterial injury in the setting of trauma, although uncommon, can have serious consequences. An understanding of these entities and their characteristic imaging findings allows for accurate diagnosis and effective treatment.

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