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VIEWPOINT

Kawasaki Disease in Adults

A Call to Action to Manage Lifelong Coronary Risk

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Kawasaki disease (KD) is an emerging and under-recognized condition that poses a diagnostic challenge for many adult cardiologists. As the most common cause of acquired heart disease in children, KD was first described in Japan in 1967 but was not widely recognized in the United States until the 1970s.[1] Furthermore, standard therapy with intravenous immunoglobulin to prevent coronary artery aneurysm (CAA) formation was not adopted until the mid-1980s.[2] Consequently, many adults who were never diagnosed or treated for KD in childhood are now presenting with acute manifestations of cardiac disease. In addition, individuals previously diagnosed and treated for KD in childhood are frequently lost to follow-up as they enter adulthood.[3] With estimates projecting that 1 in 1,600 U.S. adults will have a history of KD by 2030, there is a strong need for adult cardiologists to have increased awareness of this vasculopathy.[4]

The prevalence of KD sequelae in adults is notable; in one U.S. study, 5% of adults aged <40 years undergoing coronary angiography for suspected myocardial ischemia had evidence of coronary sequelae from KD.[5] Moreover, a Japanese registry of 798 adults hospitalized for severe sequelae of KD found that 19.7% presented with an acute coronary syndrome (ACS) and 53.1% presented with heart failure or arrhythmias.[6] These events clustered in a bimodal pattern, peaking in young adulthood (<20 years) and again in the fourth decade of life. Moreover, many adults with KD and CAAs do not receive optimal care. In the Japanese registry, nearly 25% of hospitalized KD survivors were active smokers, and only 25% and 31% were discharged on an oral anticoagulant and statin, respectively.[6]

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The pathophysiology of KD is fundamentally different from atherosclerosis. In KD, an acute, necrotizing arteritis destroys the elastic lamina, with replacement of the media by proliferating myofibroblasts and dense fibrotic tissue.[7,8] In vivo studies using virtual histology-intravascular ultrasound confirm that unlike lipid-laden atherosclerotic plaques, the dominant component of KD lesions is fibrotic tissue.[9] For this reason, the primary risk with CAAs in KD is not aneurysm rupture but in situ

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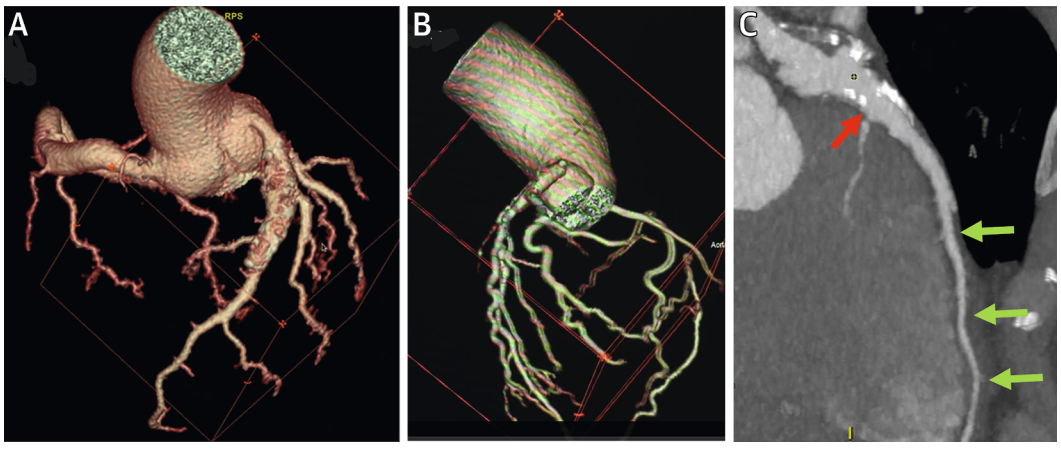
FIGURE 1 Computed Tomography of Kawasaki Disease Aneurysms



(A) Three-dimensional view of a coronary computed tomography angiogram (CTA) from a 48-year-old patient with Kawasaki disease (KD) in childhood revealing bilateral giant aneurysms with diffuse calcifications in the left anterior descending artery. (B) Coronary CTA from a 54-year-old patient with a history of KD and complete total occlusion of large aneurysms in the left main coronary artery and left anterior descending artery with a rich network of collateral vessels. (C) Coronary CTA depicting focal proximal calcifications only (red arrow) consistent with KD, with a normal distal vessel without coronary calcium (green arrows).

thrombosis. Computer modeling of flow dynamics within these aneurysms reveals a profoundly prothrombotic environment characterized by low wall shear stress, high particle residence time, and an increased oscillatory index.[10]

KD should be suspected in patients with ACS who demonstrate angiographic evidence of large, proximal CAAs or stenoses with relatively normal distal vessels. Differentiation from other causes of CAA, most commonly atherosclerosis and more rarely other vasculitides, connective tissue diseases, and infectious causes, should be considered.[11] For all such patients, a careful history is warranted, including interviewing the patient's parents, if feasible, to inquire about a prolonged illness with high fever in childhood.[5]

When adults present for follow-up with a known history of KD but earlier medical records and imaging studies are not available, two risk stratification pathways can be considered. First, a computed tomography (CT) calcium score and stress imaging study may be pursued. Coronary artery calcification in KD usually reflects organized and layered mural thrombus within old CAAs rather than atherosclerotic plaque.[7] The negative predictive value of a calcium score has not been validated in KD. However, a high coronary calcium score in the proximal coronary arteries in the absence of traditional risk factors for atherosclerotic cardiovascular disease should prompt coronary CT angiography (CTA) to identify CAAs (Figure 1).[12,13] Alternatively, a first-line coronary CTA may be performed to identify obstructive or nonobstructive coronary aneurysms. For patients with ACS, intravascular ultrasound is essential to evaluate the true size of the vessel lumen, which can be obscured by a large thrombus burden.

Management must be tailored to past and current coronary artery architecture. A recent American Heart Association Scientific Statement recommends that patients with large or giant aneurysms (absolute diameter >8 mm or Z-score ≥10) should be treated with an oral anticoagulant in conjunction with antiplatelet therapy.[14] Statin therapy should be instituted for its lipid-lowering and anti-inflammatory effects; counseling for traditional risk factors for superimposed atherosclerotic disease is critical.[15] For patients without limiting symptoms or evidence of myocardial ischemia, medical management is preferred over interventional approaches, with monitoring ideally performed by stress echocardiography to minimize radiation exposure in those who require serial testing.

Percutaneous treatment of coronary lesions in patients with KD has recognized challenges. Because KD-related lesions are often fibrotic and calcified, rotational atherectomy and/or intravascular lithotripsy may be required for adequate lesion preparation before coronary stenting.[16] Furthermore,

intravascular ultrasound or optical coherence tomography is particularly important to enable optimal stent sizing within CAAs. With coronary artery bypass graft surgery, greater degrees of competitive flow within grafted vessels can increase the risk for bypass graft failure.[17] Consequently, surgical revascularization should be reserved for patients without percutaneous treatment options, with a strong preference given to the use of arterial grafts because of the limited long-term patency of saphenous vein grafts.[18] For patients with complete total occlusion of a coronary artery, medical management is preferred in those without limiting symptoms or evidence of myocardial ischemia, because extensive collaterals often develop in childhood (**Figure 1**).[19]

Finally, there is a strong need to better connect pediatric and adult cardiologists through formal Health Care Transition programs.[20] Disrupted follow-up is associated with significantly higher in-hospital mortality.[6] Although the optimal "medical home," within Adult Congenital Heart Disease programs, adult cardiology, or shared care, is debated, the principles of transition remain fundamental. Effective transition must prioritize data portability across electronic health systems and may be spurred by triggers such as pregnancy, insurance status, or the need for specialized ACS pathways. Ensuring a "warm handoff" with a clear medical summary, transition coordinators, and a guideline-directed management plan is vital to improving long-term outcomes.

In summary, KD with CAA is a lifelong condition that requires vigilant identification and management by adult cardiologists. Improved outcomes for this vulnerable population depend on the implementation of evidence-based medical therapies, specialized transition programs, and a high index of suspicion in young adults presenting with proximal coronary lesions. For more detailed information regarding recognition and management of adults with CAA after KD in childhood, you are invited to watch a video.[21]

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