Kawasaki disease: surgical treatment.

Fabio Navarro[1] and Leonardo Mulinari [2]

[1]Hospital Pequeno Principe
[2]University of Miami Miller School of Medicine

September 19, 2022

Abstract

Kawasaki Disease (KD) is a systemic vasculitis of small and medium arteries, preferably affecting coronary arteries. It is one of the most frequent causes of acquired heart disease in children. Despite being comprehensively studied, its etiopathogenesis is not totally explained. The surgical procedures usually become necessary during the late follow-up and may be coronary artery bypass grafting, cardiac defibrillator implantation with or without cardiac resynchronization therapy, or cardiac transplantation.

Title

Kawasaki disease: surgical treatment.

Authors and affiliations

Fabio Binhara Navarro, MD

Cardiothoracic Surgery

Hospital Pequeno Principe

Leonardo Mulinari, MD, PhD

Associate Professor of Surgery

Department of Surgery

University of Miami

Corresponding author

Leonardo Mulinari, MD, PhD

1611 NW 12th Ave

East Tower Suite 3016

Miami, FL, USA 33136-1015

Kawasaki disease: surgical treatment.

Abstract

Kawasaki Disease (KD) is a systemic vasculitis of small and medium arteries, preferably affecting coronary arteries. It is one of the most frequent causes of acquired heart disease in children. Despite being comprehensively studied, its etiopathogenesis is not totally explained. The surgical procedures usually become necessary during the late follow-up and may be coronary artery bypass grafting, cardiac defibrillator implantation with or without cardiac resynchronization therapy, or cardiac transplantation.

Kawasaki Disease (KD) is a systemic vasculitis of small and medium arteries, which preferably affects coronary arteries. It is one of the most frequent causes of acquired heart disease in children. Despite being comprehensively studied, its etiopathogenesis is not totally explained. Probably genetic susceptibility allows an abnormal immunological and inflammatory reaction triggered by a viral or bacterial infection. Besides the fact of being described for more than 50 years, diagnostic confirmation is a clinical challenge. This happens because its symptoms may be similar to bacterial or viral infections and, mainly because there is no confirmatory laboratory test. The diagnosis of the typical KD is made by the presence of fever for more than five days in association with four of the five following signs and symptoms: bilateral conjunctival edema or inflammation; changes in lips or oral mucosa; polymorphic exanthema; cervical lymphadenopathy; edema, erythema, or desquamation in hands or feet. Frequently, several cardiovascular complications may occur, including valvulitis, myocarditis, pericarditis, and even a circulatory shock (KD Shock Syndrome). Coronary artery aneurysms (CAAs) and dilatation of them are more often in the subacute phase, where up almost 20% of untreated children may develop aneurysms.[1-5]

The KD treatment is essentially medical, based on intravenous immunoglobulins and aspirin in the acute phase. There are other therapeutic options like corticosteroids, plasma exchange, TNF-a inhibitors, cyclosporine, cyclophosphamide, and methotrexate. Patients with giant coronary aneurysms ought to receive double platelets anti-aggregation therapy and anticoagulation with warfarin or LWFH to prevent coronary thrombosis. The earlier treatment, preferably within the first 10 days of the disease, may assure a lower risk of CAA and therapeutic failure. Regardless of the early and adequate therapy, 8% of the children may exhibit cardiovascular complications, 6% with coronary dilatation, 1% with CAA, and 0,13% with giant CAA. The long-term management begins 4 to 6 weeks after the fever onset when the acute symptoms have already disappeared and the coronary involvement reached the maximum extent. At this moment, the treatment objective is to prevent coronary thrombosis and myocardial ischemia. Even after the resolution of the inflammatory process and luminal diameter regression, the coronary artery wall structure and function may be abnormal, leading to progressive stenosis and occlusion over time. The acute coronary occlusion treatment is done with thrombolytic agents and, if the patient has adequate size can be done percutaneously. Patients with moderate coronary aneurysms ([?]6mm) present a higher risk of developing coronary progressive occlusion or Acute Coronary Syndrome. However, this propensity increases exponentially in patients with large aneurysms (luminal diameter Z value [?] 10 or [?] 8mm), demanding a close follow-up.[4, 6-9]

The surgical procedures usually become necessary during the late follow-up. As Wang, Z et al have described in this issue of the JCS, surgical interventions may be necessary for treating myocardial ischemia when there is no possibility of percutaneous coronary intervention (PCI). When the patient has cardiomyopathy, due to coronary occlusion and myocardial ischemia or secondary to the initial myocardial insult during the acute KD, cardiac defibrillator implantation with or without cardiac resynchronization therapy, or cardiac transplantation may be necessary.

When coronary artery bypass grafting (CABG) is necessary, the precise comprehension of the myocardial ischemia mechanism and myocardial viability evaluation is very important, since it can be due to coronary stenosis or secondary to recurrent thrombosis of coronary aneurysms. In the discussed article, this mechanism is not precisely described, but, in clinical practice, it is mandatory, requiring complementary exams like myocardial scintigraphy, coronary tomography, magnetic resonance, and/or coronary angiography, preferable associated with IVUS and FFR.[7,10]

The CABG has excellent results in KD and, likewise adults, the*in-situ* artery grafts have better long-term patency in children. In addition, these arterial grafts have a growing potential that may follow the somatic child's growth, which does not happen with saphenous vein grafts. Hence, the elective choice of saphenous vein grafts, even using the no-touch technic, needs more scientific validation, mainly in smaller children.[7,11]

Albeit not found in the discussed manuscript, valvular lesions may occur and require surgical alleviation. During the acute phase, in rare situations (1,5% of cases), mild or moderate mitral insufficiency can happen, secondary to valvulitis or sub-valvar apparatus damage. Generally, this insufficiency diminishes after the resolution of the inflammatory state. Likewise, ascending aorta inflammation and dilatation of the aortic

root have also been described, leading to secondary aortic insufficiency. Mitral or aortic valvar repair or replacement have already been reported, despite being very uncommon.[7, 12, 13]

1. Kawasaki T. Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children. *Allergy* . 1967; 16: 178–222.

2. Singh S, Jindal AK, Pilania RK. Diagnosis of Kawasaki Disease.*Int J Rheum Dis.* 2018; 21: 36–44.

3. Ayusawa M, Sonobe T, Uemura S, et al. Revision of diagnostic guidelines for Kawasaki disease (the 5th revised edition). *Pediatr Int.* 2005; 47 (2), 232–4.

4. McCrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a scientific statement for health professionals from the American heart association.*Circulation* 2017; 135: e927–99.

5. Agarwal S, Agrawal DK. Kawasaki Disease: Etiopathogenesis and Novel Treatment Strategies. *Expert Rev Clin Immunol* . 2017; 13(3): 247–258.

6. Galeotti C, Bayry J, Kone-Paut I, Kaveri SV. Kawasaki disease: Aetiopathogenesis and therapeutic utility of intravenous immunoglobulin.*Autoimmun Rev.* 2010; 6 (9): 441–8.

7. Fukazawa R, Kobayashi J et al. JCS/JSCS 2020 Guideline on Diagnosis and Management of Cardiovascular Sequelae in Kawasaki Disease.*Circ J.* 2020; 84: 1348–1407.

8. Manlhiot C, Millar K, Golding F, McCrindle BW. Improved classification of coronary artery abnormalities based only on coronary artery z-scores after Kawasaki disease. *Pediatr Cardiol* . 2010; 31:242–249.

9. Gidding SS, Shulman ST, Ilbawi M, Crussi F, Duffy CE. Mucocutaneous lymph node syndrome (Kawasaki disease): delayed aortic and mitral insufficiency secondary to active valvulitis. *J Am Coll Cardiol* . 1986; 7: 894–897.

10. Ogawa S, Ohkubo T, Fukazawa R, et al. Estimation of myocardial hemodynamics before and after intervention in children with Kawasaki disease. *J Am Coll Cardiol.* 2004; 43**:** 653 – 661.

11. Kitamura S, Seki T, Kawachi K, et al. Excellent patency and growth potential of internal mammary artery grafts in pediatric coronary artery bypass surgery: New evidence for a “live” conduit. *Circulation.*1988; 78**:** I129 – I139.

12. Mishima A, Asano M, Saito T, Yamamoto S, Ukai T, Yoshitomi H, Mastumoto K, Manabe T. Mitral regurgitation caused by ruptured chordae tendineae in Kawasaki disease. *J Thorac Cardiovasc Surg.* 1996; 111: 895–896.

13. Fuse S, Tomita H, Ohara T, Iida K, Takamuro M. Severely damaged aortic valve and cardiogenic shock in an infant with Kawasaki disease.*Pediatr Int* . 2003; 45:110–113.