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CASE REPORT
Cardiovascular imaging

Multimodal imaging for cardiac follow-up and management of giant coronary aneurysm related to Kawasaki disease in childhood: a case report

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Background Kawasaki disease (KD) is a rare paediatric condition that can lead to giant coronary aneurysms. Follow-up of such complex coronary lesions remains a challenge, and their management is difficult to standardize.

Case summary Our present case concerns a 17-year-old boy who suffered a giant aneurysm of the left coronary artery, complicated by an asymptomatic stenosis. During regular follow-up, his annual cardiopulmonary exercise test revealed signs of ischaemia (ST depression and premature ventricular complexes). After several further stress tests with inconsistent results, he underwent invasive coronary angiography that revealed significant stenosis with a positive fractional flow reserve (FFR).

Discussion We discuss the challenges of diagnosing and managing coronary artery stenosis in paediatric patients with KD, particularly in cases with calcified and thrombosed lesions. A multimodal approach is crucial, including non-invasive imaging, and coronary angiography with optical coherence tomography and FFR. The evaluation of the lesion and its follow-up is an important factor in anticipating the best therapeutic choice for each patient.

Keywords Giant aneurysms • Kawasaki disease • Coronary artery bypass grafting • Case report

ESC curriculum 3.1 Coronary artery disease • 2.1 Imaging modalities

Learning points

- Kawasaki syndrome can lead to severe chronic coronary lesions that require a dedicated follow-up.
- Complex coronary lesions should be investigated using different types of non-invasive stress tests that can include CMR, echocardiography, and scintigraphy.
- An anatomic coronary imaging is recommended every 1–5 years. Regarding its accuracy with calcified lesion (optical coherence tomography) and the complementary functional information (fractional flow reserve), coronary angiography remains the first-choice exam.

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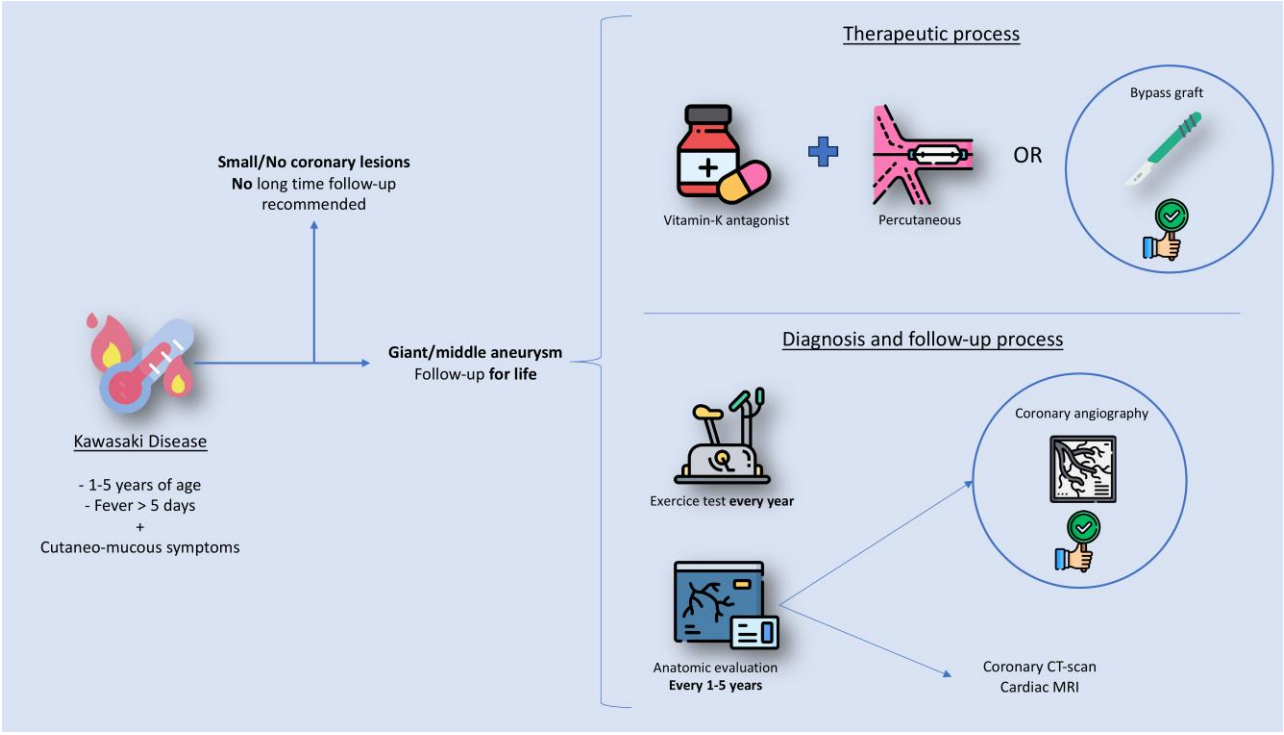
Introduction

Kawasaki disease (KD) mainly affects children under the age of 5. This inflammatory disease results in necrotizing vasculitis of small- and medium-sized arteries. Its severity is related to the onset of cardiac complications, especially coronary artery aneurysms.[1] These aneurysms usually appear during the first 6 weeks after the initial symptom and can lead to permanent giant aneurysms (GAs), defined as having a diameter >8 mm or +10 *Z*-score.[2] The mortality rate of KD fell from 2% in the 70s to 0.01% nowadays, due to high-dose immunoglobulin therapy and early anti-thrombotic prophylaxis. There is a greater risk of thrombosis or stenosis in GA, leading to acute or delayed myocardial ischaemia. ESC 2019 guidelines highlighted the value of multimodal imaging in the assessment of the severity of chronic coronary syndrome in the adult population.[3] However, there are no specific guidelines for the diagnosis and management of chronic coronary artery lesions in the paediatric population with KD.

We present here the case of a 17-year-old boy presenting with a GA of the left coronary artery complicated by severe stenosis. We discuss the management of this case, and the benefits to decision-making offered by a more accurate imaging modality.

Summary figure



Case presentation

Kawasaki disease was diagnosed when the boy was 7 months old. In the first weeks following the diagnosis, a GA of the proximal part of the left anterior descending artery (LAD) occurred. A combined antiplatelet (aspirin) and anticoagulation (Vitamin K Antagonist) therapy was conducted. However, thrombosis and stenosis of the LAD aneurysm developed over time, as observed by systematic coronary computed tomography (CT) angiography.

The patient remained asymptomatic, without clinical symptoms or signs of ischaemia in regular stress tests performed throughout childhood. At the age of 17, his annual cardiopulmonary exercise test revealed a decreased aerobic capacity, an ST segment depression in lateral leads, and premature ventricular complexes at peak effort, without any simultaneous symptom.

Therefore, complementary exams were conducted to confirm myocardial ischaemia: the coronary CT angiography underscored a worsening of the proximal LAD stenosis at the level of a voluminous calcified plaque downstream of the 7 mm aneurysm (*Figure 1*). The myocardial stress scintigraphy was negative; the coronary angiography showed a decreased fractional flow reserve (FFR) at 0.69 in front of the LAD stenosis. The optical coherence tomography (OCT) illustrated the persistence of the GA of the proximal LAD up to the first diagonal artery division, and a proximal stenosis (stenosis fraction 72%) associated with a stenosis of the diagonal (hence a bifurcation lesion) (*Figures 2* and *3*). Finally, stress cardiac magnetic resonance imaging (CMR) confirmed reversible ischaemia in the LAD territory (*Figure 4*).

The surgical team decided to perform a double coronary artery bypass grafting using a 'Y-assembly' with the right internal mammary artery connected to the post-aneurysm LAD segment and the left intern mammary artery connected to the first diagonal. The patient did not present any post-surgical complications. One month later, he engaged in a post-operative rehabilitation programme with benefit as he regained a normal physical capacity with a normalization of the ECG.

Figure 1 Coronary CT angiography image: multiplanar reformation shows left ventricle and stretched left anterior descending artery (LAD) with proximal aneurysm and calcified stenosis.

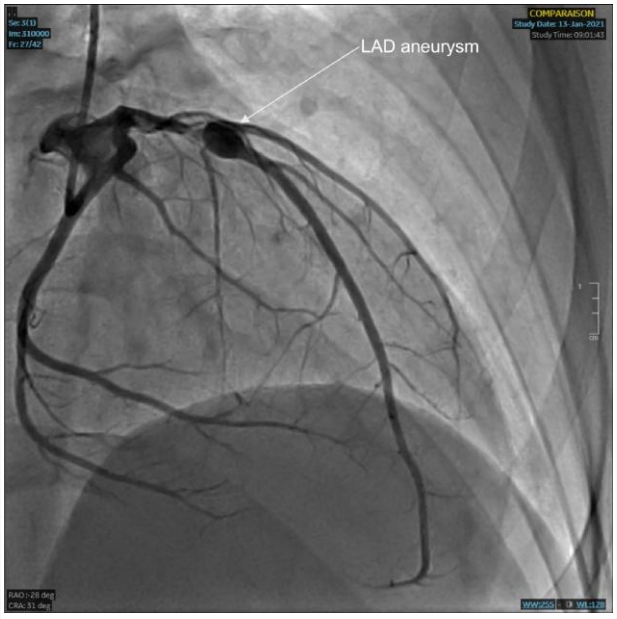


Figure 2 Coronary angiography in a projection right anterior oblique 30°/cranial 30° shows the LAD aneurysm with sub-occlusive stenosis and the absence of lesion of other coronary vessels. LAD, left anterior descending artery.

A repeated coronary angiography at 6-month post-operatively showed an evolution of the stenosis of the LAD (90–99%), a visual decrease in the GA size, and no competitive flow between the native coronary and the bypass graft.

The 9-month post-operative cardiopulmonary exercise test showed a normalized aerobic capacity without any significant cardiological anomaly.

Discussion

Giant aneurysm related to KD is rare (0.13%), but is associated with a greater risk of thrombosis or stenosis leading to myocardial ischaemia.[2] These heterogeneous vascular lesions evolve due to calcified and thrombosed mechanisms, and challenge both the diagnosis of myocardial ischaemia and its management. While ~50% of patients will experience a coronary stenosis and/or an ischaemic event during the 15 years following a KD, the prediction of a significant myocardial ischaemia remains difficult. Therefore, lifelong personalized and regular follow-up is highly recommended.[2]

In this case, a positive cardiopulmonary exercise test led to the performance of a coronary CT angiography. This showed a worsening of the LAD stenosis. However, the anatomic evaluation of a GA with extended calcified lesions can be limited by the use of non-invasive imaging, given that calcified lesions can lead to blooming artefacts. Regular coronary angiography appears to be one of the best ways to assess the level of stenosis, extension, and stability of coronary lesions.[4] Intravascular coronary imaging, which includes intravascular ultrasound (IVUS) and OCT, is a cornerstone tool in the description of coronary parietal lesions (atherosclerosis, intimal hyperplasia, calcification). This is recommended for the diagnostic and therapeutic processes offered to KD patients where intravascular intervention is required. The technical means most adapted to paediatric patients would appear to be OCT, given that its spatial resolution is 10 times higher than IVUS.[5,6] In this case, it showed specific signs of KD lesions (intimal hyperplasia and calcified lesions.[6]) That said, OCT also highlighted the underestimated focal stenosis of the initial part of the first diagonal, which was included in the GA. This information was crucial in evaluating which type of therapeutic plan could be considered for this patient.

Evaluating inducible ischaemia to warrant risk stratification is recommended in the follow-up of KD. AHA guidelines recommend the reasonable use, prior to any interventional procedure, of non-invasive stress tests, such as the stress scintigraphy that was performed in our case.[7] Since 2012, ESC guidelines regarding adults with chronic coronary diseases recommend coronary angiography to evaluate the haemodynamic significance of a lesion with the FFR.[3] This invasive examination is still considered a second-line test. However, the safety of coronary angiography should expand its use in paediatrics, as proposed by the 2020 JCS guidelines.[2] The advocated cut-off value is 0.8 in the adult population. The cut-off value in the paediatric population would appear to be closer to 0.75.[2] Our case underscores the value of a positive FFR result, suggesting a need for a second ischaemic test (cardiac stress MRI) to validate the recommendation for coronary revascularization, even after a previously normal myocardial scintigraphy.

The type of coronary revascularization in this case merits discussion, as it was a single vessel lesion.[2] Different percutaneous intervention techniques are available to treat KD coronary stenosis as follows: percutaneous transluminal coronary rotational ablation, stenting, and angioplasty. However, the long-term result depends on the patient's age and body size, and the number and locations of the coronary stenoses.[8] Percutaneous intervention techniques also present a higher risk of complication in the paediatric population, because of a lack of dedicated catheters.[9] In this case, the pre-operative assessment advocated for a surgical coronary artery bypass graft, given the complexity of LAD stenosis and its high level of obstruction, which constitute a positive criterion for the long-term patency of bypass grafts with no risk of competitive flow.[10] Lastly, the anatomic coronary angiography overview supported this therapeutic choice. Indeed, the significant focal stenosis of the diagonal, in addition to the LAD stenosis, supported the case for an upgrade to a double coronary artery bypass graft.[11]

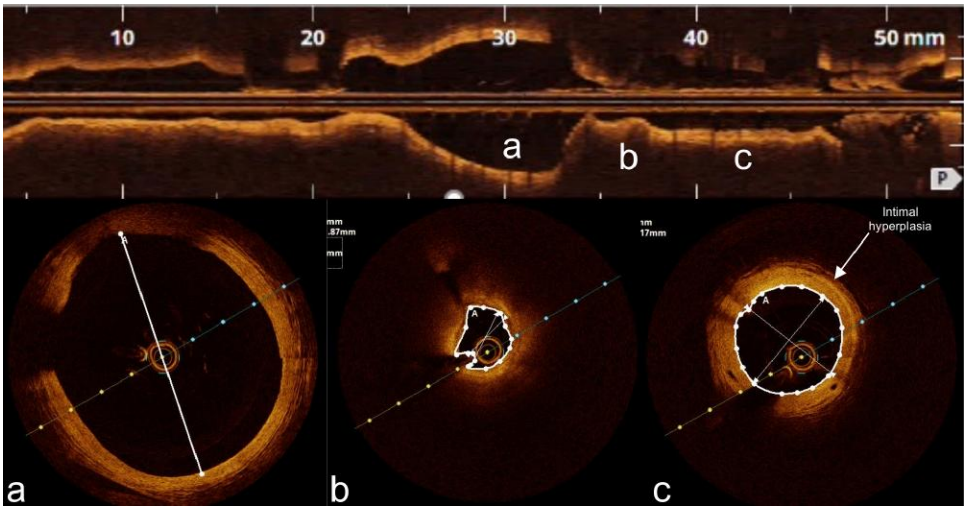


Figure 3 Coronary optical coherence tomography (OCT) of the LAD obtained during the coronary angiography; longitudinal view with corresponding axial views: (*A*) LAD aneurysm measured as 7.2 mm maximal diameter. No evidence of intra-aneurysm thrombus. Moderate intimal hyperplasia. (*B*) Calcified stenosis of the LAD aneurysm corresponding to a 77% stenosis. (*C*) Significant intimal hyperplasia (arrow) following the LAD aneurysm. Then, the coronary artery returns to a normal diameter (3 mm).

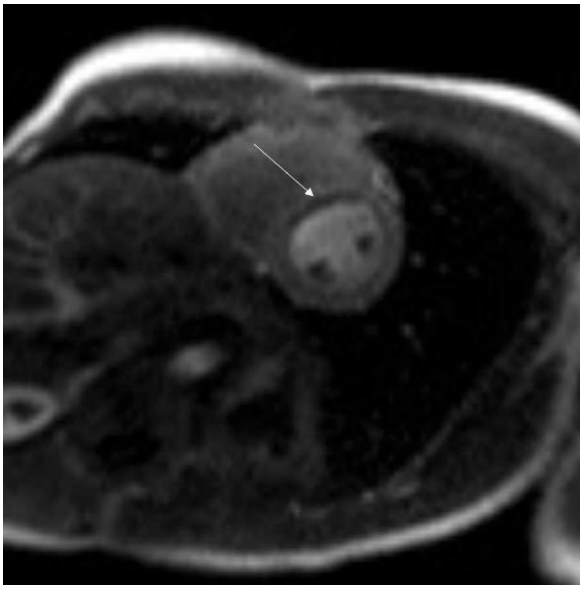


Figure 4 Stress CMR, myocardial first pass perfusion image of a basal short axis view showing a delay of perfusion (arrow) at the level of the basal septal wall, corresponding to the LAD territory.

Conclusion

We have presented the case of an asymptomatic 17-year-old child with a stenosis of a giant coronary aneurysm related to KD. Several examinations were performed to confirm the indication of revascularization and its modality, highlighting the usefulness of anatomical (OCT) and functional (FFR) coronary angiography in decision-making regarding complex and heterogeneous coronary lesions.

Lead author biography

Paediatric cardiologist in the University Hospital of Montpellier with a specialization in cardiac imaging.

Consent: This case report was written with patient consent and is in accordance with COPE guidelines.

Conflict of interest: None declared.

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Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.

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