



Asian Pacific Society of Cardiology Consensus Statement on Management of Coronary Artery Disease in Adults with Late Complications of Kawasaki Disease

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Abstract

Despite reductions in the mortality of Kawasaki disease with advances in its treatment, a proportion of patients still develop coronary artery dilatation that may persist even into adulthood. These lesions carry an increased risk of myocardial ischaemia and infarction. However, published clinical guidelines on the management and long-term follow up of patients with these late complications of Kawasaki disease are limited. The Asian Pacific Society of Cardiology convened an expert panel to review the available literature and develop consensus recommendations to guide clinicians in this area. The panel developed statements on the assessment and risk stratification of coronary artery disease, investigations for follow up, as well as considerations around treatment of stenotic lesions with medical therapy, percutaneous coronary intervention and coronary artery bypass grafting. Each statement was voted on by each panel member and consensus was reached when 80% of experts voted 'agree' or 'neutral'. This process resulted in the development of consensus recommendations to guide cardiologists and internists in the follow up and management of patients with coronary artery lesions as a late complication of Kawasaki disease.

Keywords

Kawasaki disease, coronary artery disease, coronary aneurysms, long-term follow up, percutaneous coronary intervention, Asia-Pacific, consensus

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Kawasaki disease, first described in 1967, is an acute, self-limiting, febrile vasculitis that predominantly affects children under the age of 5 years. Treatment comprises primarily aspirin and intravenous immunoglobulin, along with corticosteroids, infliximab, other immunosuppressants and adjunctive plasmapheresis for refractory cases. Even with treatment, cardiovascular complications may arise, including coronary artery dilatation, valvular lesions, pericarditis and myocarditis. Patients who do not fulfil four of the five principal clinical features of Kawasaki disease, but are found to have coronary artery abnormalities, are deemed to have Kawasaki disease, albeit an incomplete form.

Almost all deaths in patients with Kawasaki disease result from its cardiovascular sequelae.³ Coronary artery dilation may progress to the formation of coronary aneurysms, which may persist into

adulthood and carry an increased risk of death, MI and the need for coronary revascularisation. Even if coronary artery lesions eventually regress, the underlying tissue is not normal, and people diagnosed with these lesions have an increased lifetime risk of cardiac events.

There are limited published clinical guidelines on the management and long-term follow-up of coronary artery disease in adults with late complications of Kawasaki disease. Therefore, the Asian Pacific Society of Cardiology (APSC) developed these consensus recommendations to provide expert guidance. These recommendations are intended to guide cardiologists and internists managing cardiovascular conditions. However, the consensus recommendations are meant to supplement – but not replace – clinical judgement.

Methods

The APSC convened a 12-member panel to review the existing literature, discuss gaps in the current management strategies, outline areas where further guidance is needed and — ultimately — develop consensus recommendations on the follow up and management of late coronary artery sequelae resulting from Kawasaki disease. The experts were mostly members of the APSC who were nominated by national societies and endorsed by the APSC consensus board, as well as international experts in treating coronary artery aneurysms. These consensus recommendations build on previous work by the American Heart Association and the Japanese Circulation Society (JCS)/Japanese Society for Cardiovascular Surgery.^{2,6}

Consensus recommendations were developed by a process similar to that outlined in the 2019 American College of Cardiology methodology for creating expert consensus decision pathways. Draft consensus recommendations were developed based on the existing literature around the management of coronary sequelae of Kawasaki disease and circulated to the expert panel. Recommendations were discussed in a consensus meeting held on 11 May 2022. The statements were then each put to an independent online vote using a three-point scale (agree, neutral, or disagree). Consensus was reached when 80% of experts voted agree or neutral.

When there was no consensus, the statements were further discussed via email then revised accordingly until the criteria for consensus were reached.

Experts also reviewed the existing literature and appraised the evidence behind each consensus statement using the Grading of Recommendations, Assessment, Development, and Evaluation system as follows:

- 1. High (authors have high confidence that the true effect is similar to the estimated effect).
- 2. Moderate (authors believe that the true effect is probably close to the estimated effect).
- 3. Low (true effect may be markedly different from the estimated effect).
- Very low (true effect is probably markedly different from the estimated effect).

Consensus Recommendations Assessment and Risk Stratification of Coronary Artery Lesions

Statement 1. Coronary artery lesions should be quantitatively assessed for intra-luminal dimensions and classified into small, medium and giant aneurysms accordingly (*Tables 1 and 2*). Level of evidence: Moderate (high 2; moderate 8; low 2). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 2. Coronary CT, cardiac magnetic resonance (MR), echocardiographic assessment and coronary angiogram are all valid methods of initial assessment of coronary artery lesion diameter. If information from multiple imaging modalities is available, the largest dimension measured should be used to classify the lesion. In adults, it is also reasonable to consider the CT result as most indicative of true aneurysm diameter.

Level of evidence: Moderate (high 2; moderate 8; low 2). Level of consensus: 91.7% agree; 8.3% neutral; 0% disagree. **Statement 3.** If available, initial Z-scores measured during the acute phase of the illness as well as maximal Z-scores measured at least 1 month after the acute phase should be considered for risk stratification of coronary artery lesions, especially in patients with coronary artery lesions that subsequently regress (*Table 3*). Level of evidence: Moderate (high 3; moderate 8; low 1).

Level of consensus: 100% agree; 0% neutral; 0% disagree.

The initial classification of coronary artery lesions is important because it has prognostic relevance for subsequent outcomes. $^{4.8}$ The presence and size of the coronary artery lesions are the most important factors for judging severity of Kawasaki disease. The JCS/JSCS 2020 guidelines suggest the use of the Z-score for evaluating the severity of coronary artery lesions by their intra-luminal diameter. 6 Use of the Z-score is strongly recommended for patients aged ≥ 5 years (*Table 1*). $^{2.6,9;10}$

However, the JCS/JSCS 2020 guidelines on this relate primarily to children, and different formulae exist. $^{2.6}$ In adults, there will patients in whom a normal segment may be ≥ 4 mm. Furthermore, the calculation of Z-scores in adults can be complicated and not practical for routine clinical practice. We propose the use of the alternative definition in the 2013 JCS guidelines, as indicated in *Table 2.* While the 2013 guidelines also apply to children, it can be more pragmatically applied to adults, as coronary artery aneurysms are sized relative to adjacent segments.

We also recognise that the absolute cut-off of 8 mm remains a useful reference point in defining giant aneurysms in the paediatric population, even if it may not be as significant in an adult population in which coronary arteries are naturally expected to be of larger diameter. Hence, while using the pragmatic method of classifying coronary aneurysms, we suggest that coronary artery aneurysms identified as ≥ 8 mm during assessment in childhood/teenage years retain the label of giant aneurysm into adulthood, unless they undergo significant regression.

It should be noted that other authors define coronary aneurysms as focal dilations of at least 1.5 times the adjacent normal segment. 12-15 Furthermore, it has also been highlighted that a standard definition of 'giant aneurysm' does not exist. 16 In the broader literature, the term 'giant aneurysm' has been accorded to aneurysms >20 mm, and this is used for aneurysms regardless of the underlying pathophysiology or cause. 12,16 These discrepancies between aneurysms in general, and aneurysms arising from Kawasaki disease, should be acknowledged as an area of uncertainty.

In asymptomatic patients known to have a prior history of Kawasaki disease, coronary CT, cardiac MR, echocardiography and angiography are all valid imaging modalities to screen for remaining coronary artery lesions. The choice of imaging tool should consider the patient's age and clinical characteristics, along with the availability of hardware and expertise in treating centres.

We recognise that each of these tools has its limitations. We also acknowledge that measurements of coronary artery lesion diameter may be complicated by calcification in the aneurysm wall. When amalgamating information from multiple imaging modalities, we suggest using the largest dimension measured to classify the coronary artery lesion. In adults, it is also reasonable to consider the CT scan result as indicative of true aneurysm diameter.

When available from childhood records, the initial Z-scores measured during the acute phase of the illness, as well as the maximal Z-score measured at least 1 month after the acute phase, should be considered in the risk stratification of coronary artery lesions. This has particular implications for patients in groups II or III (*Table 3*), where the previous presence of aneurysmal lesions that persist more than a month after the acute phase (as opposed to transient dilatation that resolves within a month) should prompt closer initial follow-up, even if the lesions do subsequently regress. ^{2,6} For selected patients, additional invasive investigation with coronary angiography may be considered to diagnose stenotic lesions within aneurysmal segment and assess for ischaemic consequences via measurement of fractional flow reserve (FFR). This classification informs the risk of long-term sequelae, such as ischaemic heart disease caused by these coronary artery lesions. ^{5,17,18} It will also be used to quide follow-up and management.

Long-term Follow-up of Coronary Artery Lesions

Statement 4. Consider interval investigations and long-term follow up of Kawasaki disease patients with coronary artery lesions as guided by their risk level (*Table 4*) except for those without coronary involvement or those with only transient coronary artery dilatation.

Level of evidence: Moderate (high 1; moderate 6; low 5). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 5. Consider stress echocardiography or a treadmill stress test as a minimally invasive method for follow-up assessment of inducible ischaemia post Kawasaki disease. Alternative forms of stress imaging (e.g. stress nuclear scan, stress MRI) may also be considered, especially in adults.

Level of evidence: Moderate (high 1; moderate 7; low 4). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 6. Coronary angiography is the gold standard investigation and should be performed in patients with evidence of inducible myocardial ischaemia investigation.

Level of evidence: High (high 8; moderate 3; low 1). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Follow-up for patients with coronary artery sequelae of Kawasaki disease should continue long-term, with interval evaluations for inducible ischaemia as well as consideration of re-imaging the coronary arteries. ^{2,6} The degree of follow up should be guided by each patient's risk level (*Table 4*).

Patients with transient dilation can be discharged from on-going cardiology care after follow-up for 5 years if luminal dimensions return to normal by 1 month. This should be clarified against aneurysms that have regressed. For regressed aneurysms, it is reasonable to follow up on Kawasaki disease patients even in the long term because of potential coronary events in adulthood and the functional and structural abnormalities in the coronary vessel wall in the long-term.

Nevertheless, all patients should be reminded that having had Kawasaki disease is part of their permanent medical history and should be highlighted to medical professionals in the future. They should be counselled on general cardiovascular health and risk factors, including leading a healthy lifestyle.

Table 1: Coronary Artery Sizing by Z-score

| Coronary Artery Lesion | Z-score |
|------------------------|--|
| No involvement | Z-score <2 |
| Dilatation only | Z-score ≥2 to <2.5 |
| Small aneurysm | Z-score \geq 2.5 to <5 (inner diameter \geq 3 mm to <4 mm) |
| Medium aneurysm | Z-score \geq 5 to <10 (inner diameter \geq 4 mm to <8 mm) |
| Giant aneurysm | Z score ≥10 (inner diameter ≥8 mm) |

Table 2: Pragmatic Coronary Artery Aneurysm Sizing

| Coronary Artery Lesion | Internal Diameter |
|------------------------------|---|
| Small aneurysm or dilatation | <1.5 times that of an adjacent segment |
| Medium aneurysm | 1.5~4 times that of an adjacent segment |
| Giant aneurysm | ≥4 times that of adjacent segment OR absolute diameter ≥8 mm when diagnosed as a child/teenager (unless there is significant subsequent regression) |

Table 3: Severity Classification

| Risk | Level | Definition | |
|------|--|---|--|
| I | | No dilation change Z-score always <2 including during the acute phase | |
| II | | Transient dilatation Mild transient dilation that normalises by 1 month after onset | |
| III | Illa: Small aneurysm during acute phase | Regression CALs persisting beyond 1 month from onset, | |
| | IIIb: Medium/giant aneurysm during acute phase | which subsequently normalise during follow-up. Do not fall into group V. | |
| IV | IVa: Small aneurysm | Remaining coronary aneurysms | |
| | IVb: Medium aneurysm | CALs that remain beyond 1 month from onset that do not fully normalise during follow-up. Do | |
| | IVc: Giant aneurysm | not fall into group V. | |
| V | Va: Without ischaemic findings | Coronary artery stenotic lesion Coronary angiography shows stenotic lesion in the coronary artery | |

CAL = coronary artery lesions; Vb = with ischaemic findings.

Patients with medium or giant aneurysms in the acute phase, as well as all those with remnant aneurysmal coronary artery lesions, should receive regular assessments for inducible ischaemia. Stress echocardiography or treadmill stress tests should be considered as minimally invasive means of assessing for inducible ischaemia. Patient forms of minimally invasive stress imaging, for example, stress nuclear scans or stress MRI, may also be considered, especially in adults.

There are additional considerations in children pertaining to these alternative imaging modalities. CT angiography provides a valuable non-invasive method of assessing coronary arteries, and a CT-derived FFR value can also be calculated, but it requires pharmacological control of heart rate and IV contrast use, as well as the risk of radiation exposure. Myocardial perfusion imaging is sensitive and useful for detecting

Table 4: Long-term Follow-up of Coronary Artery Lesions

| Risk I | Level | ECG, Echocardiogram | Assessment for Inducible Ischaemia | Coronary Imaging (CT, MRI, CAG) |
|--------|-------------------------------------|--|------------------------------------|--|
| I | No dilation | Assess at 1, 2, 6, 12 months, | Not necessary | Not necessary |
| II | Transient dilation | and 5 years (or yearly) until 5 years old | | |
| Illa | (Acute phase) small aneurysm | Yearly | Not necessary | Consider at convalescent phase, 1 year from onset, or when the aneurysm regresses. Recommended on finishing high school. |
| IIIb | (Acute phase) medium/giant aneurysm | Every 6–12 months | Consider every 3–5 years | Consider at convalescent phase, 1 year, then every 3–5 years |
| IVa | Remaining small aneurysm | Yearly | Consider every 3–5 years | Consider at convalescent phase, 1 year, then every 3–5 years |
| IVb | Remaining medium aneurysm | Every 6–12 months | Consider every 2–5 years | Consider at convalescent phase, 1 year, then every 2–5 years |
| IVc | Remaining giant aneurysm | Every 6–12 months | Consider every 1–5 years | Consider at convalescent phase, 1 year, then every 1–5 years |
| Va | Stenotic lesion without ischaemia | Every 6–12 months | Consider yearly | Consider at convalescent phase, 1 year, then every 1–5 years |
| Vb | Stenotic lesion with ischaemia | Consider timely follow-up | Consider timely follow-up | Consider timely follow-up |

CAG = coronary angiography.

myocardial ischaemia but carries the risk of radiation and the risk of drug load needed to provide myocardial stress. Cardiac MRI uses no radiation, but requires control of heart rate. It also has a long imaging duration, thus requiring a long sedation time.

In patients where an initial minimally invasive assessment reveals evidence of inducible myocardial ischaemia, coronary angiography should be performed. This enables detailed imaging of the coronary artery lumen and is the gold standard for evaluating the degree of stenosis, prognosticating the lesion and deciding if therapeutic intervention is indicated. Patients with stenotic lesions that are functionally significant and cause myocardial ischaemia (*Table 4*, group Vb) require closer attention and follow up. Hence, we suggest that the treating clinician exercises discretion in deciding the interval for follow-up investigations.

Medical Therapy for Coronary Artery Lesions

Statement 7. Low-dose aspirin should be continued for all patients with remaining aneurysmal coronary artery lesions of any diameter (risk level IV and above).

Level of evidence: High (high 6; moderate 4; low 2). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 8. Dual antiplatelet therapy (i.e. aspirin + a $P2Y_{12}$ inhibitor) may be considered in patients with remaining medium or giant aneurysms.

Level of evidence: Low (high 1; moderate 2; low 9). Level of consensus: 91.7% agree; 8.3% neutral; 0% disagree.

Statement 9. Warfarin, in combination with low dose aspirin, should be administered to patients with remaining giant aneurysms, a history of MI or known thrombosis in the coronary artery lesion, with a target international normalised ratio (INR) of 2.0–3.0. Level of evidence: Moderate (high 1; moderate 11; low 1). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 10. Triple therapy with aspirin, additional antiplatelet and warfarin can be considered in patients with high risk for thrombosis; for example, those with giant aneurysms and recent history of coronary artery thrombosis or stent implantation.

Level of evidence: Low (high 1; moderate 3; low 8). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 11. Direct oral anticoagulants (DOACs) may be considered as an alternative to replace warfarin in the recommendations above. However, it should be noted that data regarding its use in this disease are limited. Nonetheless, given the body of evidence in other areas of anticoagulation/thromboembolic disease, such as AF, deep vein thrombosis, pulmonary embolism and left ventricular (LV) thrombus, the use of DOACs in place of warfarin may be considered after appropriate discussion with patients.

Level of evidence: Low (high 0; moderate 2; low 10). Level of consensus: 91.7% agree; 8.3% neutral; 0% disagree.

Statement 12. Empirical statin therapy can be considered in patients with remaining aneurysms of any size for their pleiotropic effects. Level of evidence: Low (high 1; moderate 3; low 8). Level of consensus: 91.7% agree; 8.3% neutral; 0% disagree.

Aspirin is already well established as standard therapy in acute treatment of Kawasaki disease. ^{29,30} We suggest that low-dose aspirin should be continued as primary prophylaxis against ischaemic heart disease in patients with remaining aneurysmal segments (risk level IV and above). ^{2,6,31}

Alternative antiplatelet agents should be considered for patients with resistance to aspirin or aspirin intolerance. In patients with remnant medium or giant aneurysms, dual antiplatelet therapy comprising aspirin plus another $P2Y_{12}$ inhibitor may be considered.

Patients with remaining giant aneurysms, a history of MI or known thrombosis in the coronary artery lesion should be anticoagulated with warfarin in addition to low-dose aspirin, with a target INR of 2.0–3.0. Warfarin therapy, in combination with aspirin, has been shown to lower the incidence of MI as compared to aspirin alone. ^{32,33} Target INR should take into consideration size of the aneurysm, clinical condition as well as the patient's bleeding risk, with a lower target INR of 2.0–2.5 in patients with higher bleeding risk. Low molecular weight heparin (LMWH) is a reasonable alternative for patients who are unable to achieve a stable level of anticoagulation on warfarin.³⁴

DOACs are a relatively new class of drugs, and there are limited data pertaining to their use in anticoagulation of giant aneurysms resulting from Kawasaki disease. Nonetheless, given the body of evidence around their use in other indications for anticoagulation, such as AF, deep vein thrombosis, pulmonary embolism and LV thrombus, they may be considered as an alternative anticoagulant to replace warfarin in the recommendations above.

Triple therapy with aspirin, an additional antiplatelet and warfarin can be considered in patients with high risk for thrombosis, such as those with giant aneurysms and recent history of coronary artery thrombosis or stent implantation.^{2,31} This can be continued for a short period of time, in line with other guidelines concerning stent implantation in patients with a separate indication for anticoagulation.³⁵

Finally, there is some suggestion that statins may have a role in reducing coronary artery inflammation via their pleiotropic effects.³⁶ Statins may thus be considered as adjunctive medical therapy in patients with remaining coronary artery lesions of any size.

Invasive Therapy for Coronary Artery Lesions

Statement 13. In acute MI, primary percutaneous coronary intervention (PCI) should be considered as primary reperfusion therapy.

Level of evidence: High (high 9; moderate 3; low 0). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 14. For patients with high thrombus burden within the aneurysm, intracoronary thrombolysis can be considered. Level of evidence: Moderate (high 1; moderate 8; low 3). Level of consensus: 91.7% agree; 8.3% neutral; 0% disagree.

Statement 15. Intravascular ultrasound (IVUS) or optical coherence tomography (OCT) may be considered to assess true luminal dimensions to guide PCI.

Level of evidence: High (high 6; moderate 4; low 2). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 16. While plain old balloon angioplasty (POBA) may be effective for localised stenotic lesions without calcification or aneurysms less than a few years after the onset of Kawasaki disease, it is not effective for lesions with severe calcification many years after the onset of Kawasaki disease.

Level of evidence: Moderate (high 4; moderate 5; low 3). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 17. Rotational or orbital atherectomy, or other adjunctive therapy (e.g. shockwave) may be required for calcified stenotic lesions.

Level of evidence: Moderate (high 5; moderate 4; low 3). Level of consensus: 100% agree; 0% neutral; 0% disagree.

Statement 18. The primary indications for coronary artery bypass grafting (CABG) as a means of revascularisation may be considered in accordance with established guidelines in the literature for standard types of coronary artery disease.

Level of evidence: Moderate (high 4; moderate 7; low 1). Level of consensus: 100% agree; 0% neutral; 0% disagree. Coronary artery lesions with functionally significant stenosis may require emergent or elective intervention. In acute MI, we recommend considering primary PCI as the primary modality of reperfusion therapy to reperfuse the ischaemic myocardium as quickly as possible. Because data are limited, management is generally extrapolated from guidelines on management of acute coronary syndrome in adults. Intracoronary thrombolysis or intravenous antiplatelet agents (e.g. abciximab, eptifibatide and tirofiban) can be considered in patients with high thrombus burden within coronary aneurysms. Intracoronary thrombolysis has been shown to be effective in reducing thrombus burden in coronary aneurysms, even up to several hours after thrombus formation, although evidence is limited to case series. 37–39

During coronary artery intervention, it is crucial to obtain an accurate picture of true luminal dimensions to guide balloon and stent sizing. Existing literature and clinical experience suggest that it is easy to underestimate true luminal dimensions and miss underlying aneurysmal distortion. 40 We recommend the use of IVUS or OCT in the assessment of luminal dimensions. 41–44

Chronic lesions are heavily fibrotic and calcified and may be difficult to expand by balloon angioplasty alone. Even if the lesion can be expanded, there is significant recoil, which limits the subsequent results. The high pressures needed to expand the lesions have been associated with formation of neo-aneurysms at the site of dilation $^{40.43}$

As such, while POBA may be effective for localised stenotic lesions without calcifications or aneurysms, less than a few years after the onset of Kawasaki disease, we do not recommend them in severely calcified chronic lesions. These may instead require rotational or orbital atherectomy or other adjunctive therapies (e.g. shockwave). ^{43–46} In target vessels in which a burr larger than 2.15 mm in diameter can be used, good patency of the vessel can be maintained by close follow-up and repeat coronary rotational atherectomy, if required. ⁴⁶

CABG has been shown to be an effective method of revascularisation in the paediatric Kawasaki disease population, with good long-term outcomes. 47–49 We recommend considering CABG for revascularisation of chronic coronary artery lesions in adults in accordance with other established guidelines in the literature for standard types of coronary artery disease.

Limitations and Future Research

There are significant limitations to these consensus statements. Kawasaki disease is a rare condition, and the prevalence of coronary sequelae has been steadily declining with improvements in diagnosis of Kawasaki disease as well as the introduction of aspirin and intravenous immunoglobulin as effective therapies in the acute phase. As such, evidence for some of these recommendations (in particular, Statements 9, 11, 12 and 13) are extrapolated from conventional adult coronary artery disease guidelines or based on case series with limited patients, some of which were published more than two decades ago.

There is currently no published evidence for the use of DOACs in the anticoagulation of Kawasaki disease patients with high-risk coronary lesions, and these recommendations are extrapolated from the use of DOACs in place of warfarin or LMWH for other indications. Ultimately, in this rare condition, treatment decisions should be individualised and determined by the managing physician with consideration for the

ndications for CABG as per Consider intracoronary quidelines for revascularisation Consider atherectomy for thrombolysis if thrombus standard types of CAD burden in aneurysm is high KAWASAKI DISEASE Use primary PCI as primary OBA likely ineffective for reperfusion therapy in AMI lesions with severe calcifications **EMERGENCY ELECTIVE** Remaining Transient dilation artery change aneurvsms **CORONARY ARTERY PERCUTANEOUS BYPASS GRAFTING** CORONARY INTERVENTION Z-score (younger patients) Pragmatic method INVASIVE THERAPY Initial assessment via coronary CT, cardiac Т MR, echocardiogram or coronary angiogram. Use the largest dimension measured. **RISK STRATIFICATION** Consider DAPT Consider triple patients with giant aneurysms, previous MI or known high-risk patients† If available, use initial Z-scores during aneurysm thrombosis acute phase and maximal scores 1 month after for risk stratification ow-dose aspirin for all patients Warfarin (INR 2.0-3.0), LMWH, DOAC with remaining aneurysms [†]For example, giant aneurysms with recent MI or stent implantation Low-risk patients do not Coronary angiography remains gold standard require lifelong follow-up Interval follow-up based on risk level, with: **MEDICAL THERAPY** FOLLOW-UP Echocardiogram Functional assessment Coronary imaging angiography, cardiac MR

Figure 1: Summary of Consensus Recommendations

AMI = acute MI; CABG = coronary artery bypass grafting; CAD = coronary artery disease; DAPT = dual antiplatelet therapy; DOAC = direct oral anticoagulants; INR = international normalised ratio; LMWH = low molecular weight heparin; MR = magnetic resonance; PCI = percutaneous coronary intervention; POBA = plain old balloon angioplasty.

patient's unique profile. These consensus recommendations are meant to serve merely as a guide and not replace clinical judgement.

Further research is needed to determine the optimal medical therapy of remnant coronary artery lesions post Kawasaki disease, specifically, which combinations of antiplatelets/anticoagulation are most efficacious in the prevention of ischaemic heart disease while balanced against bleeding risks at each level of risk.

Conclusion

A summary of the consensus recommendations is shown in *Figure 1*. Advances in the management of Kawasaki disease have significantly reduced its mortality. Nevertheless, a proportion of patients may have residual coronary artery aneurysms that do not regress after the acute illness, which increases the risk of MI. We have developed these consensus recommendations to guide risk stratification, long-term follow up and management of coronary artery disease in adults with these late complications of Kawasaki disease.

Clinical Perspective

- Coronary artery lesions resulting from Kawasaki disease should be assessed for their intra-luminal dimensions and risk stratified accordingly.
- Risk level guides choice of investigations and intensity of long-term follow-up.
- Medical management for coronary artery lesions includes aspirin and statin, along with additional antiplatelet agents and/or anticoagulation in higher-risk patients.
- Primary percutaneous coronary intervention should be considered as primary reperfusion therapy in acute MI, with consideration of intracoronary thrombolysis or intravenous antiplatelet if the thrombus burden is high.
- During intervention, the use of intravascular ultrasound or optical coherence tomography for accurate luminal sizing and atherectomy for calcified lesions should be considered.

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