

Review



가와사키병에서 관상동맥 병변의 중재술 및 항혈전 전략: 지침 기반 관리

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Catheter-Based Revascularization and Antithrombotic Strategies for Post-Kawasaki Disease Coronary Artery Lesions: A Narrative Review of Guideline-Aligned Management

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Abstract

Coronary artery aneurysms, stenosis, and thrombosis following Kawasaki disease (KD) remain clinically significant from childhood through adulthood, producing ischemia driven by complex vascular remodeling rather than atherosclerosis. As survivors age, revascularization decisions become frequent and complex, demanding consideration of unique features like ectasia, intimal hyperplasia, and calcification. This narrative review synthesizes contemporary evidence and guideline frameworks [American Heart Association (AHA) 2017/2024; Japanese Circulation Society (JCS) 2020] to clarify appropriate catheter-based revascularization strategies. We summarize risk stratification guided by lesion morphology and long-term prognosis. Pre-procedural evaluation integrates coronary computed tomography angiography for anatomic mapping with adjunctive intravascular ultrasound or optical coherence tomography to characterize calcium and aneurysm-neck geometry. In anatomically complex or aneurysmal vessels where traditional physiology may be misleading, PET-derived myocardial flow reserve can provide complementary insight. For focal, technically approachable stenosis, percutaneous coronary intervention (PCI) is feasible when performed with imaging guidance and tailored lesion preparation. Conservative rotational atherectomy may be required for heavy calcification, and stent selection must accommodate abrupt caliber transitions around aneurysms. Covered stents, such as PK Papyrus, are reserved strictly for acute perforation. In contrast, diffuse multivessel disease, left-main involvement, or aneurysm-complex anatomy generally favors coronary artery bypass grafting (CABG), supported by favorable long-term outcomes in contemporary Japanese series. In these settings, CABG—typically with an internal thoracic artery-first strategy—provides durable graft patency and remains the preferred revascularization approach. Antithrombotic therapy requires individualized balancing of thrombosis and bleeding risk. Aspirin and dual antiplatelet therapy are applied in usual PCI contexts, while giant aneurysms or prior thrombosis often warrant warfarin-based anticoagulation, with limited but emerging adult data on selective direct oral anticoagulants (DOACs) use. Ultimately, KD-related

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coronary disease requires imaging-guided, physiology-informed, and anatomy-specific strategies. A heart-team approach and structured longitudinal follow-up remain essential for optimizing outcomes as these patients transition into adulthood.

Keywords: Kawasaki Disease; Coronary Aneurysm; Coronary Stenosis; Percutaneous Coronary Intervention; Coronary Artery Bypass; Ultrasonography, Interventional; Tomography, Optical Coherence; Fractional Flow Reserve, Myocardial; Myocardial Perfusion Imaging; Anticoagulants

Introduction

Kawasaki disease (KD) is an acute vasculitis with persistent coronary sequelae—luminal dilation/aneurysm, mural thrombosis, and progressive stenosis driven by myofibroblastic intimal hyperplasia and calcification [1–3]. Patients with medium or large coronary artery aneurysms (CAA) remain at risk for late ischemic events despite timely IVIG [1,3,4]. As KD survivors enter adulthood, clinicians face revascularization decisions whose biomechanics and risks differ from atherosclerotic coronary artery disease (CAD) [5].

This review offers stepwise, practical guidance for catheter-based interventions across the KD lifespan.

Main Subject

1. Literature search and guideline basis

We conducted a narrative review (2000–October 2025) using PubMed/EMBASE/Cochrane with the terms: KD, coronary aneurysm, percutaneous coronary intervention (PCI), rotational atherectomy (RA), covered stent, PET myocardial flow reserve, American Heart Association (AHA) guideline, Japanese Circulation Society (JCS) guideline. We prioritized AHA (2017; 2024 update) [1,3] and JCS (2020) guidance [2], pediatric/adult congenital heart disease catheterization consensus (2024) [6], and KD-specific series on direct oral anticoagulants (DOACs), RA, and coronary artery bypass grafting (CABG) outcomes.

2. Natural history and pathology of Kawasaki disease coronary lesions

Late coronary sequelae of KD remain clinically distinct from adult atherosclerotic disease. The presence of large or giant CAA, typically defined as having a maximal diameter of ≥ 8 mm or z-score ≥ 10 , is a critical predictor of adverse events, including thrombosis, stenosis, myocardial infarction, and the subsequent need for reintervention [4,7]. Stenosis characteristically develops at the proximal and distal segments of the aneurysms, a process driven by chronic vascular remodeling and fibrous intimal thickening rather than plaque buildup. Furthermore, calcification is a common pathological finding, often becoming evident years after the acute phase [4,5,7,8]. These unique pathological realities—aneurysm geometry, intimal thickening, and calcification—significantly elevate the importance of precise

intravascular imaging [specifically intravascular ultrasound (IVUS) or optical coherence tomography (OCT)] and necessitate a conservative approach to stent sizing and landing-zone selection during interventional procedures.

3. Risk stratification frameworks

The clinical management of post-KD coronary disease is primarily guided by risk stratification frameworks from major international societies. Both the AHA and the JCS center their classifications on the coronary artery z-score.

The AHA 2017 statement established five distinct risk levels based on the maximum and current z-scores, including sub-levels for regressed aneurysms. The subsequent 2024 update further refined this stratification by providing clearer definitions of acute-phase high-risk criteria—such as age \leq 6 months, elevated inflammatory markers, and coronary artery z-scores ≥ 2.5 in the left anterior descending (LAD) or right coronary artery (RCA)—and by modifying the recommended long-term follow-up schedule [1,3]. The acute-phase high-risk criteria in the AHA 2024 statement include age \leq 6 months, elevated inflammatory markers, and coronary artery z-scores ≥ 2.5 in the LAD or RCA. The JCS 2020 guidelines utilize z-score categories (small, medium, giant) alongside a classification of long-term status (I–V) that places particular emphasis on the presence or absence of myocardial ischemia and stenosis [2] (Table 1).

4. Indications and strategies for revascularization

The decision to pursue revascularization in patients with KD is guided by the presence of symptoms and objective evidence of ischemia within the affected coronary territories. Indications for intervention include symptomatic angina or the documentation of objective ischemia through stress imaging or PET-derived myocardial flow reserve (PET-MFR). Furthermore, revascularization is warranted in cases of left ventricular dysfunction associated with reversible ischemia, progressive critical stenosis [particularly in the left main (LM) or proximal LAD arteries], recurrent thrombosis/embolization, or complex, high-risk anatomy such as bilateral ostial disease [1–3]. The choice between PCI and CABG is dictated by the complexity and morphology of the coronary lesions. PCI is generally favored for focal, technically approachable stenoses or when ideal surgical targets are lacking, such as in the setting of diffuse ectasia. CABG remains the preferred strategy for managing diffuse multivessel disease, complex left-main involvement, or other forms of aneurysm-complex anatomy. The use of CABG is supported by data from Japanese national KD cohorts, which

Table 1. Comparative summary of long-term risk stratification frameworks in KD: AHA 2017/2024 vs. JCS 2020

Variables	AHA 2017 [1]	AHA 2024 [3]	JCS 2020 [2]
Classification	5 levels (1–5) by max/current z-score; sub-levels for regressed aneurysm	Defines acute-phase high-risk (e.g., age \leq 6 months or LAD/RCA z ≥ 2.5); clarifies follow-up cadence	z-score categories (small/medium/giant) + long-term status I–V emphasizing ischemia/stenosis
Antithrombotic emphasis	Escalate ASA/anticoagulation with risk level; giant CAA \rightarrow ASA + warfarin/LMWH	Selective DOAC consideration in adult giant CAA	Similar intensity rules; anticoagulation in giant CAA

Practical implication: Both guidelines center on z-scores; AHA sharpens high-risk; JCS adds long-term status.

KD: Kawasaki disease; AHA: American Heart Association; JCS: Japanese Circulation Society; LAD: left anterior descending artery; RCA: right coronary artery; ASA: aspirin; CAA: coronary artery aneurysm; LMWH: low-molecular-weight heparin; DOAC: direct oral anticoagulant.

report low 30-day mortality and excellent 10-year survival rates [9–11]. Specific indications and the comparative roles of PCI versus CABG are comprehensively presented in Table 2. A detailed comparison of PCI and CABG—including anatomic suitability, procedural risks, long-term outcomes, and practical considerations—is summarized in Table 3.

5. Pre-interventional evaluation: integrating anatomy and physiology

Optimal outcomes for PCI in post-KD lesions necessitate a comprehensive evaluation that integrates high-resolution anatomical imaging with functional physiological assessment. Anatomical assessment is initiated with coronary computed tomography angiography (CTA), which is crucial for comprehensive mapping of aneurysm/ectasia, the distribution of calcification, and the location of side-branch takeoffs. Subsequently, IVUS or OCT is mandatory to precisely define key lesion characteristics, including the calcium arc and length, and complex aneurysm-neck geometry relevant for safe stent sizing and optimal landing zones [8].

For physiological assessment, traditional Fractional Flow Reserve measurements may be confounded in aneurysmal or serial lesions due to flow separation and increased vascular capacitance. Therefore, PET-MFR is considered a more reliable measure to reflect the true ischemic burden in post-KD vasculopathy [12]. In settings of complex anatomy, physiological data serve as a complementary tool to guide interventional decision-making.

6. Percutaneous coronary intervention for stenosis: a stepwise technical approach

The interventional strategy for KD-related stenosis must be tailored to the unique non-atherosclerotic characteristics of the vessel wall, namely intimal hyperplasia and calcification.

Procedural planning requires the use of 6–7 appropriate French guide catheters, with care

Table 2. Key clinical and anatomic indications for revascularization: PCI versus CABG

Clinical/anatomic situation	Suggested first-line
Symptomatic/objective ischemia in focal, accessible lesion(s)	PCI
Left-main (esp. proximal) or bilateral ostial high-risk stenosis	CABG preferred (PCI bridge if single, discrete lesion)
Stenosis at giant CAA neck / suboptimal stent landing zones	CABG ± selective PCI adjunct (heart-team)
Progressive stenosis or recurrent thrombosis despite optimal therapy	Individualized (PCI or CABG)
Acute PCI complications (perforation)	Emergency PCI; covered stent bail-out

PCI: percutaneous coronary intervention; CABG: coronary artery bypass grafting; CAA: coronary artery aneurysm.

Table 3. Comprehensive comparison of therapeutic domains, outcomes, and limitations for PCI versus CABG

Domain	PCI	CABG
Typical indication	Focal, technically approachable stenosis; poor surgical targets; bridge to surgery	Diffuse multivessel or left-main disease; aneurysm-complex anatomy
Main advantages	Less invasive; rapid recovery; preserves future options	Arterial graft patency; comprehensive treatment of complex proximal disease
Main limitations	Malapposition risk across ectatic/aneurysmal segments; reintervention; calcification may require RA	Sternotomy; peri-operative risks; growth issues in children
Long-term signals	KD RA series: event-free survival ≈ 79% at 10 y; ≈ 39% at 20 y (small samples) [16]	National survey (Japan, 2008–2019): 30-day mortality 0.9%; 10-year survival 94% [11]
Practical tip	IVUS/OCT optimization; distal-reference sizing; use PET-MFR for complex physiology	ITA-first strategy; heart-team planning

PCI: percutaneous coronary intervention; CABG: coronary artery bypass grafting; RA: rotational atherectomy; IVUS: intravascular ultrasound; OCT: optical coherence tomography; PET-MFR: positron emission tomography-derived myocardial flow reserve; ITA: internal thoracic artery.

taken to avoid deep seating near aneurysms or ostia to prevent localized injury. Plain balloon angioplasty (PBA) may be employed for initial focal dilation or to facilitate subsequent device crossing, especially in small children or when stenting is deferred. However, PBA carries a high risk of elastic recoil and restenosis, which limits its use as a definitive monotherapy in fibrotic KD lesions [13–15]. In addition, coronary balloon angioplasty carries a potential risk of neoaneurysm formation—particularly when high-pressure balloons are used—because mechanical stress may disrupt the structurally fragile arterial wall remodeled after KD.

Lesion preparation is paramount for achieving adequate stent expansion. In cases of heavy calcification (defined by arc > 180°, length > 5 mm, or predicted under-expansion), RA is recommended, supported by KD-specific RA outcomes [16]. RA should be performed conservatively, utilizing low burr-to-vessel ratios, employing short runs, and strictly followed by post-atherectomy intravascular imaging. Because no clear consensus exists between drug-eluting stent (DES) and bare-metal stent (BMS) choices in KD patients, stent selection is typically individualized based on bleeding risk and concurrent antithrombotic therapy.

The stent strategy involves sizing the stent to the distal reference vessel diameter, and securing long landing zones that extend beyond aneurysmal transitions. Final optimization must be performed under IVUS/OCT guidance, carefully avoiding aggressive oversizing. When anatomical imaging and physiological data are discordant, functional assessment with PET-MFR should be incorporated to guide the decision to intervene [12].

Alternatively, intravascular lithotripsy has recently emerged as an effective tool for circumferential and nodular calcium fracture, offering a less ablative option that may be particularly beneficial in vessels with irregular geometry typical of post-KD coronary arteries [17]. Both techniques require post-atherectomy/lithotripsy intravascular imaging to confirm adequate plaque modification. A comparative overview of balloon angioplasty, stent implantation, RA, and intravascular lithotripsy is provided in Table 4.

Table 4. Technical comparison of percutaneous interventional options for KD-related stenosis

Variables	Balloon angioplasty	Stent implantation	Rotational atherectomy	Intravascular lithotripsy
Primary goal	Focal dilatation/lesion preparation (including cutting/scoring)	Scaffold + drug release to suppress restenosis	Modify/ablate calcium to enable delivery/expansion	Fracture deep/circumferential calcium to facilitate stent expansion/delivery
Advantages in KD	No implant → preserves growth/future surgery	Immediate lumen gain; restenosis suppression	Enables adequate stent expansion in heavy calcification	Lower risk of distal embolization/slow flow compared to RA; effective for deep calcium fracture
Main limitations	Elastic recoil/dissection; higher mid-term restenosis in calcific/aneurysm-adjacent segments	Malapposition risk across ectatic/aneurysmal transitions; DAPT required	Slow-flow/microembolization; perforation; caution near aneurysm neck	Requires balloon access; limited data in pediatric KD
Indication	Very focal, non-calcific stenosis or preparation	Straight, focal lesions with good landing zones; IVUS/OCT-guided optimization	IVUS/OCT shows arc > 180°/length > 5 mm or predicted under-expansion	Severe, deep, or nodular calcification; when RA is not feasible or carries high risk
Antithrombotics	Usually aspirin alone (context-dependent)	DAPT 6–12 months (shorter if HBR); minimize triple therapy if OAC needed	Per final device (eg. DES, typically requiring DAPT)	Per final device (often DES → DAPT)
Evidence notes	Case series/registries	Adult CAD adapted to KD; imaging optimization decisive	Small KD series: event-free survival ≈ 79% at 10 y; ≈ 39% at 20 y	Emerging data; primarily adult CAD adapted; case reports in refractory KD lesions

KD: Kawasaki disease; DAPT: dual antiplatelet therapy; DES: drug-eluting stent; IVUS: intravascular ultrasound; OCT: optical coherence tomography; OAC: oral anticoagulation; HBR: high bleeding risk; CAD: coronary artery disease.

7. Treating aneurysm-related problems

Management of complications specific to KD-related aneurysms requires highly specialized strategies. In the event of acute coronary perforation during PCI, a covered stent (e.g., the PK Papyrus) is indicated under humanitarian device exemption as a bail-out measure requiring immediate hemodynamic surveillance [18].

It is crucial to note that elective aneurysm exclusion is not an approved indication for covered stents. For a saccular side-branch aneurysm associated with a thromboembolism risk but supplying minimal myocardial territory, stent-assisted coil or plug embolization remains a niche option requiring meticulous protection, given the limited supporting evidence [19,20]. For large or giant proximal aneurysms complicated by diffuse distal disease, CABG generally remains the preferred primary revascularization strategy, with selective endovascular adjuncts considered only after multidisciplinary heart-team discussion [9–11].

8. Antithrombotic therapy

Antithrombotic management in patients with post-KD lesions requires a careful, individualized balancing of thrombosis risk (due to aneurysms or stents) and bleeding risk.

Following stent implantation, dual antiplatelet therapy (DAPT) is typically prescribed for 6–12 months. A shorter duration may be considered for patients deemed high bleeding risk [21,22]. When oral anticoagulation (OAC) is required concurrently, efforts should be made to minimize triple therapy (OAC + two antiplatelet agents), preferably utilizing a simplified regimen of OAC plus clopidogrel [21,22]. For Giant CAA or prior thrombosis (defined as ≥ 8 mm or z-score ≥ 10), the regimen generally combines low-dose aspirin with warfarin. The target international normalized ratio (INR) is typically maintained between 2.0–2.5 but may be escalated to 2.5–3.0 based on individual risk assessment [1–3,23,24]. When a temporary interruption of warfarin is necessary, such as for surgery, bridging anticoagulation with low-molecular-weight heparin (LMWH) is indicated. In the adult population with giant CAA, DOACs may be considered selectively, provided there is strict specialist oversight; however, evidence supporting their routine use in the pediatric and young adult KD populations remains limited [3,25].

Table 5 provides a summary of these antithrombotic regimens, aligning them with patient risk stratification and device context.

9. Peri-procedural management in pediatrics and young adults

Interventional and imaging procedures in the pediatric and young adult KD population necessitate special attention to safety and developmental considerations.

1) Sedation and monitoring

Sedation and monitoring for pediatric and adolescent patients should adhere to consensus guidelines, such as those published by the American Academy of Pediatrics/American Academy of Pediatric Dentistry (AAP/AAPD 2019) [6].

Table 5. Antithrombotic regimens for post-KD coronary lesions stratified by risk level

Variables	Suggested regimen	Key references
Risk Level 1 (no involvement)	Stop aspirin after acute phase (6–8 wk) if stable	[1]
Risk Level 2 (dilation only)	Low-dose aspirin short-term; discontinue if normalized	[1–3]
Risk Level 3 (small CAA, z-score 2.5–5)	Long-term aspirin; consider dual therapy if high-risk features	[1]
Risk Level 4 (medium CAA, z-score 5–10)	Long-term low-dose aspirin; add clopidogrel for persistent or current medium CAA per AHA recommendations; consider anticoagulation if stasis or thrombosis risk is present.	[1–3]
Risk Level 5 (giant CAA \geq 8 mm or z-score \geq 10)	Aspirin + warfarin (typical INR 2.0–2.5 or 2.5–3.0); LMWH bridge when interrupting	[1–3,23,24]
Adult giant CAA—DOAC	Selective consideration with specialist oversight; pediatric evidence limited	[3,25]
After DES	DAPT 6–12 mo (shorter if HBR); if OAC needed, minimize triple therapy; prefer OAC + clopidogrel	[21,22]

AHA: American Heart Association; CAA: coronary artery aneurysm; INR: international normalized ratio; LMWH: low-molecular-weight heparin; DAPT: dual antiplatelet therapy; DOAC: direct oral anticoagulant; DES: drug-eluting stent; HBR: high bleeding risk; OAC: oral anticoagulation.

2) Radiation safety

Radiation safety remains a critical concern, mandating the strict application of the as low as reasonably achievable (ALARA) principle during all fluoroscopy-guided procedures.

3) Pregnancy-capable adolescents

For pregnancy-capable adolescents and young adults, the American College of Obstetricians and Gynecologists (ACOG) guidance stipulates that clinically necessary imaging should not be withheld [26]. When feasible, non-ionizing modalities such as magnetic resonance imaging or ultrasound should be preferred, and when X-ray or computed tomography (CT) is required, the radiation dose must be minimized [27].

Conclusion

Coronary artery disease related to KD requires management strategies that extend beyond those routinely applied to atherosclerotic PCI. Successful long-term outcomes, encompassing both symptom control and sustained safety, are predicated on several key principles. These include thoughtful selection between PCI and CABG, utilization of imaging-guided lesion preparation and stenting, conservative use of covered stents strictly as a bail-out measure for acute complications, and implementation of disciplined, individualized antithrombotic management. Ultimately, a mandatory heart-team approach and vigilant, structured follow-up remain essential to optimize care as this unique patient population transitions into and progresses through adulthood.

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