



360° PEDIATRICS

IAP PEDIATRIC PRACTICE GUIDELINES

IAP Presidential Action Plan 2026

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Kawasaki Disease

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Kawasaki Disease

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Introduction

- Kawasaki disease (KD) is an acute systemic medium vessels vasculitis predominantly affecting infants and children below 5 years.
- First described in 1967 by Dr. Tomisaku Kawasaki, it has since become the leading cause of acquired heart disease.
- Pathogenesis remains an enigma, but recent advances suggest a complex interplay of immune reaction, triggered by infectious agents or environmental factors in a genetically susceptible individual.

Criteria for Kawasaki Disease

Criteria for Complete Kawasaki Disease

- **Fever for at least 4 days**
- **AND at least 4 of 5 of the following:**
 - Rash
 - Bilateral non-exudative bulbar conjunctivitis
 - Oral mucosal changes (red, cracked lips; "strawberry tongue")
 - Extremity changes (red, swollen hands and feet, or peeling skin on fingertips/toes later in the illness)
 - Unilateral cervical lymphadenopathy.

Criteria for Incomplete Kawasaki Disease

- **Fever of at least 5 days (or 7 days in infants younger than 6 months)**
- **PLUS**, at least 2-3 principal clinical features
- **AND** evidence from laboratory tests or echocardiogram:

Laboratory Findings

The presence of at least 3 of 6 criteria:

- Anaemia for age
- Platelet count $\geq 450,000/\text{mm}^3$ after the 7th day of fever
- Serum albumin ≤ 3.0 g/dL
- Elevated alanine transaminase
- WBC count $\geq 15,000/\text{mm}^3$
- Urine WBCs $\geq 10/\text{high-power field}$.

Echocardiogram Findings

If laboratory criteria are not met, an echocardiogram can be diagnostic if it shows:

- Coronary artery Z-score of ≥ 2.5 (Table 2)
- Coronary artery aneurysm plus decreased ventricular function, mitral regurgitation, or pericardial effusion.

Computed Tomography Coronary Angiography

- Indicated with giant coronary aneurysms, for thrombus, stenosis and calcifications, better delineation of distal lesions and the left circumflex artery.

Complications

- Coronary artery lesions (CALs) with aneurysmal dilation, thrombosis, and/or stenosis, leading to myocardial infarction and death
- **KD shock syndrome** (KD with poor perfusion, shock, or drop in SBP $>20\%$), often associated with severe laboratory markers of inflammation and higher risk of coronary artery anomalies (CAA)
- **Macrophage activation syndrome.**

Investigations

- Laboratory Investigations: Haemoglobin, complete blood count (CBC), platelet count, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), N-terminal pro-B-type natriuretic peptide (NT-proBNP), antistreptolysin O (ASO), liver function tests (LFTs), electrolytes, ferritin, and routine urine examination (RE)
- Other Investigations: Abdominal ultrasonography, ECG, and echocardiography.

Management

Aim in the acute phase is to control inflammation and prevent coronary artery anomalies (CAA).

- **Dosage: Intravenous immunoglobulin (IVIG)** (2 g/kg over 12 hours) with Aspirin (30 mg/kg/-day in 3-4 divided doses till fever subsides, followed by a maintenance dose of 3-5 mg/kg). though some centres favour initiation with 3-5 mg/kg
- In the absence of CAA, aspirin should be continued for 6-8 weeks
- Administer IVIG as soon as diagnosis is made, preferably within 10 days from onset of fever
- *In late diagnosis*, IVIG should still be administered if fever persists or with persistently raised CRP, ESR, or CAAs
- Even with IVIG infusion, transient coronary dilatation may occur in approximately 20% of patients that resolves within 3-6 months
- 5% of patients develop CAA, and 1% develop giant aneurysms, and therapy needs to be intensified with drugs like Infliximab/ steroids/ Cyclosporine or a combination of all.
- Patients with CAAs must receive antithrombotic therapy
 - Coronary ectasia Z score > 2: Aspirin (3-5 mg/kg)
 - Coronary ectasia Z score > 5: Additional Clopidogrel (dose 0.2 - 1 mg/kg) or Dipyridamole (1 mg/kg)
 - Coronary ectasia Z score > 10: Add low molecular weight heparin (1 mg/kg/ day subcutaneously) or warfarin. The recommended therapeutic goal is a 0.5–1.0 activated factor Xa level for LMWH or an International Normalized Ratio (INR) of 2–2.5 for warfarin
- *IVIG resistance (10- 15 %)* is defined as no defervescence by 48 hours of IVIG completion or disease recrudescence within 2 weeks.

Treatment Options

Infliximab (Preferred Option): 10 mg/Kg single infusion over 2 hrs.

- Usually, rapid defervescence and lowering of CRP by 24 to 48 hours
- Prior tuberculosis screening is not necessary

Second dose IVIg 2 gm/kg. May not work in 30% of patients.

Pulse methyl-prednisolone: 30 mg/kg/dose for 1-3 days;

Other treatment options:

- **Cyclosporin-a:** 3-8 mg/kg/ body weight /day PO
- **Anakinra:** 2-6 mg/Kg/day by subcutaneous injection x 5 days
- **Plasma exchange:** reserved when all known medical therapies
- **IVIg and steroids combination:** Used in the following cases
 - Suspected IVIG resistance according to risk scores
 - Coronary aneurysms at diagnosis
 - Complicated by macrophage activation syndrome
 - Pre-emptively for therapy intensification in infantile KD
 - Kawasaki Disease Shock Syndrome

Kawasaki Disease Shock Syndrome

- More likely to be IVIG resistant and more likely to require additional anti-inflammatory treatment.

Long-Term Sequelae and Risks

- More than 90% of small CAAs resolve over time, regression rates for large aneurysms remain lower (ranging between 26% and 35%)
- Young adults with a history of KD with normal coronary arteries are more likely to experience cardiovascular symptoms compared to controls of similar age and sex.

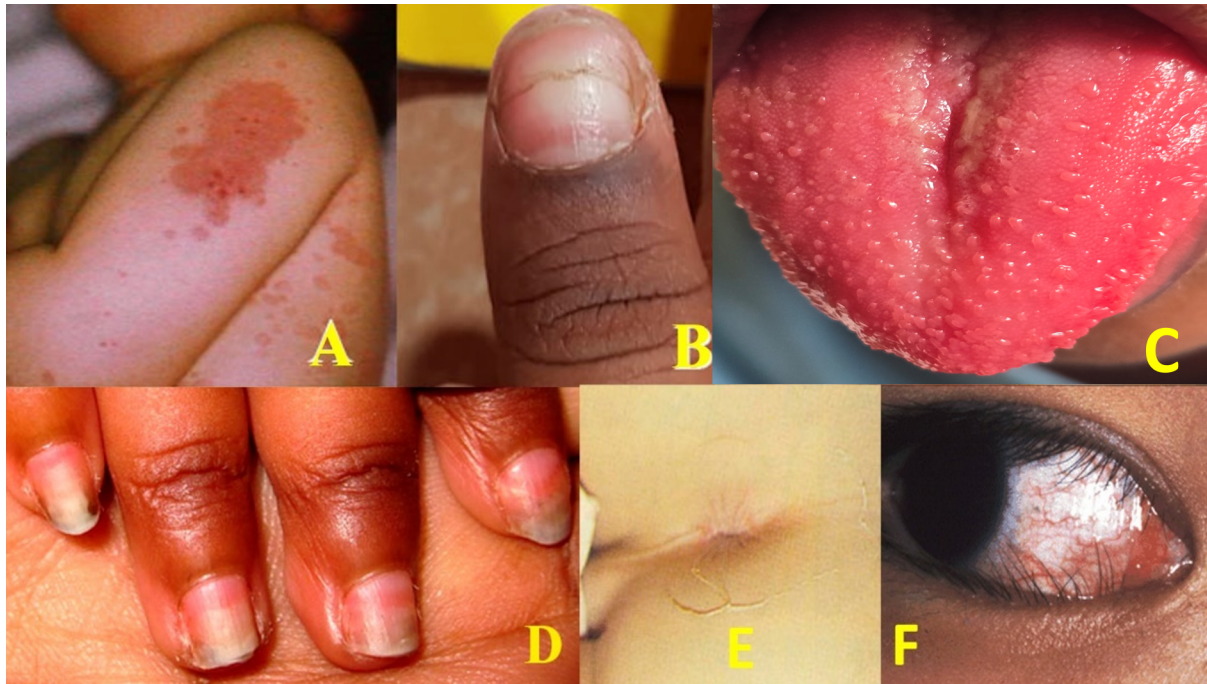
Important Considerations

- Echocardiogram should be performed at diagnosis, repeated at 1–2 weeks and 4–6 weeks, regardless of the initial findings (Figure 1)
- Diagnosis can be made with a fever of at least 4 days in the presence of 4 or more principal clinical features
- All other potential causes for the symptoms must be ruled out
- High irritability and persistent fever, especially in children less than 6 months of age, erythema around the BCG scar (18-24%), perianal excoriation, orange chromonychia on the nail bed, etc., are additional supporting evidence for the confirmation of KD.

Figure 1 A-E. Clinical signs in Kawasaki disease: **A.** Non purulent conjunctivitis **B.** Redness of lip, **C.** Erythematous maculo-papular rashes predominantly present on the trunk, **D.** Unilateral cervical lymphadenopathy, **E1&2.** Typical periungual desquamation, **F.** Swelling and redness of soles



Figure 2 A-F. Clinical & supportive evidence in KD. **A.** Erythema around the BCG scar, **B.** Beau's line- a transverse line or groove well seen over the thumb nail after 3-4 week, **C.** Strawberry tongue, **D.** Chromonychia-Striking brownish orange colour change of nails in acute phase. **E.** Perianal excoriation, **F.** Nonexudative, sparing of limbus, without exudate is an important and specific clinical sign



BCG: Bacillus Calmette–Guérin; KD: Kawasaki disease

Table 1. Major differential diagnosis

- Measles (may be associated with carditis but no coronary involvement)
- Scarlet Fever (late immunological complication: rheumatic heart disease; coronaries are spared)
- Systemic Juvenile Idiopathic Arthritis (rare transient dilatation of coronary arteries)
- Stevens-Johnson Syndrome
- Drug Reactions
- Toxic Shock Syndrome
- Rickettsial infections (rare coronary artery involvement)
- Staphylococcal Scalded Skin Syndrome
- Multi System Inflammatory Syndrome (MIS-C)(Coronary dilatation seen)

Table 2. Classification of coronary artery aneurysms and their follow-up

Coronary artery involvement	Z score/ diameter	Timing of coronary 2-D Echocardiography
No involvement	<2	1-week interval till discharge, 1month & 6month, 2-years to see intimal dysfunction
Dilatation	2 to 2.5z	1-2 week interval, 1month after treatment, 6month, then till resolving to normal
Small aneurysm	2.6 to 5 z ($\leq 5\text{mm}$)	2-week interval till no progression 1month, 6 th month and then till resolving to normal, after 2 years of stopping ASA
Medium	5 to 10z (5-8mm)	2-week interval till no progression 1month, 6 th month and then till resolving to normal, after 2 years of stopping ASA
Large/Giant	>10z (>8mm)	weekly till one month and then monthly till 3rd month, see for thrombus then every 6months plus ECG/ TMT for older children if needed/ MDCT

Figure 3. The American Heart Association guidelines regarding the long-term follow-up of Kawasaki disease starting from 1 year of age

LONG TERM FOLLOW-UP of KAWASAKI DISEASE				
RISK LEVEL 1 Normal Coronary artery: - ASA not beyond 6-8 weeks - Physical exercise – no restriction beyond 8 weeks; - yearly review, No invasive investigation.	RISK LEVEL 2 - Transient coronary ectasia or CAA - ASA not beyond 6-8 weeks - Physical exercise – no restriction beyond 8 weeks 3-5 yearly review - No invasive investigation	RISK LEVEL 3 - Small medium CAA ASA 3-5mg/kg/d till CAA regresses. - Physical exercise – No restriction in children <9 years of age beyond 6-8 weeks; - For > 9 years of age, Consider stress exercise test twice a year. - ECG, echo every year; Avoid high impact sports till antiplatelets on. Angiography and myocardial perfusion scans. - If ischemia is detected in cardiac evaluation.	RISK LEVEL 4 - Large/multiple/compl ex CAA(Z score +10) ASA /Warfarin/LMWH - till CAA regresses(increase incidence of bleeding) - Physical exercise – void high impact sports (avoid bleeding.) - Stress exercise test twice a year, ECG, echo every year; - Angiography if ischemia is suggested in CV evaluation; and repeat it if required and Myocardial perfusion scans.	RISK LEVEL 5 - Coronary artery stenosis: - ASA/Warfarin/LMW H - Investigate with stress tests, myocardial perfusion, echo/ECG twice a year or more frequent; - Angiography whenever required and plan a therapeutic intervention.

Suggested Reading

- Singh S, Kawasaki T. Kawasaki disease in India: lessons learnt over the last 20 years. *Indian Pediatr.* 2016;53(2):119-124. doi:10.1007/s13312-016-0804-5
- Singh S, Jindal AK, Pilania RK. Diagnosis of Kawasaki disease. *Int J Rheum Dis.* 2018;21(1):36-44. doi:10.1111/1756-185X.13224
- Jone PN, Tremoulet A, Choueiter N, et al. Update on diagnosis and management of Kawasaki disease: a scientific statement from the American Heart Association. *Circulation.* 2024;150(23):e481-e500. doi:10.1161/CIR.0000000000001295
- Van Stijn D, Planken RN, Groenink M, Blom N, de Winter RJ, Kuijpers T, Kuipers I. Practical workflow for cardiovascular assessment and follow-up in Kawasaki disease based on expert opinion. *Front Pediatr.* 2022;10:873421. doi:10.3389/fped.2022.873421.
- Lao F, Robinson CH, Borovsky D, Ewusie J, Beattie K, Batthish M. Long-term cardiovascular outcomes and mortality following Kawasaki disease: a systematic review and meta-analysis. *Paediatr Child Health.* 2025;30(5):406-413. doi:10.1093/pch/pxaf003
- Pilania RK, Jindal AK, Guleria S, et al. An update on treatment of Kawasaki disease. *Curr Treat Options Rheumatol.* 2019;5:36-55. doi:10.1007/s40674-019-00115-z

