Kawasaki Disease (KD)

Clinical Revision Sheet

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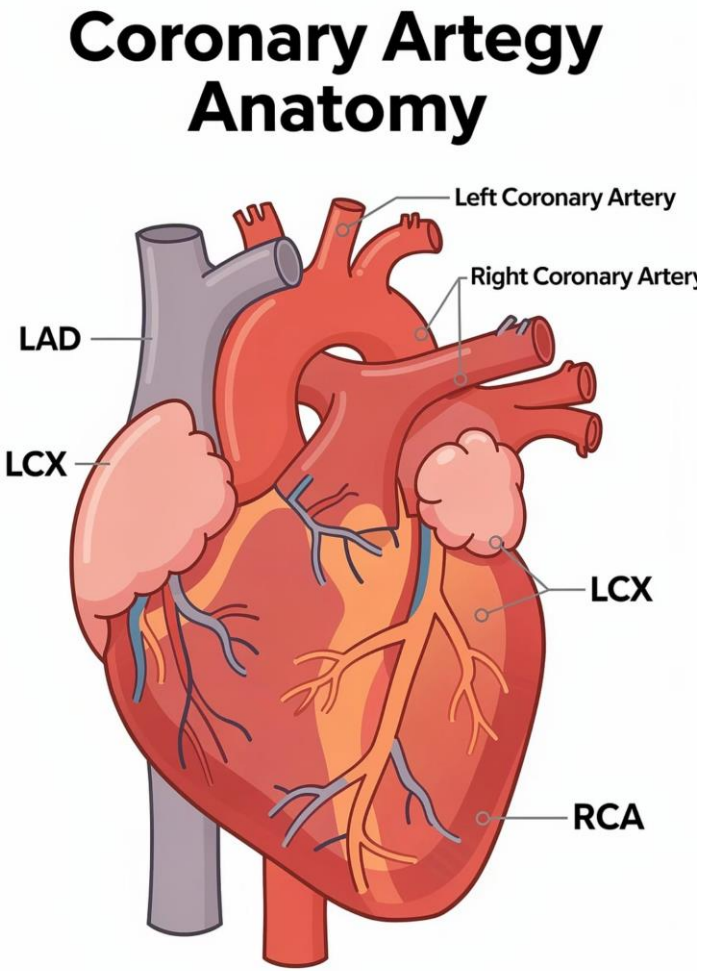


1. Definition

Kawasaki disease (KD) is an acute, self-limited systemic vasculitis of medium-sized arteries, predominantly affecting children <5 years of age. It is the leading cause of acquired heart disease in children because it can cause coronary artery aneurysms (CAA).

Clinical Pearl: Early diagnosis and treatment with IVIG within the first 10 days markedly reduce coronary artery complications.

2. Etiology

The exact cause is unknown. KD is thought to result from an abnormal immune response to an infectious trigger in genetically susceptible children, leading to systemic vasculitis, especially involving the coronary arteries.

3. Who Is at Risk?

- **Age <5 years (peak 2–3 years)**
- **Male sex**
- **Asian ethnicity**
- **Infants (<1 year) → higher risk of incomplete KD and coronary aneurysm**

4. Diagnosis

Diagnostic Criteria (Classic KD)

Fever ≥5 days PLUS ≥4 of the following 5 criteria:

Clinical Criteria	Typical Findings
Bilateral conjunctivitis	Non-purulent, painless
Oral changes	Cracked lips, strawberry tongue, red oral mucosa
Polymorphous rash	Diffuse, non-vesicular
Extremity changes	Erythema/edema of hands & feet → periungual desquamation
Cervical lymphadenopathy	≥1.5 cm, usually unilateral

Incomplete Kawasaki Disease

Suspect if:

- Fever ≥5 days
- Only 2–3 clinical criteria
- Elevated inflammatory markers
- Especially infants <6 months

⚠ ➞ Perform echocardiography immediately.

5. Clinical Features

Acute Phase (Days 1–10)

- Persistent high fever
- Bilateral non-purulent conjunctivitis
- Cracked lips
- Strawberry tongue
- Diffuse polymorphous rash
- Swollen/red hands and feet
- Cervical lymphadenopathy
- Marked irritability

Other Findings

- Vomiting
- Abdominal pain
- Diarrhea
- Sterile pyuria
- Arthritis
- Myocarditis
- Pericarditis

Subacute Phase (Weeks 2–4)

- Fever resolves
- Periungual desquamation
- Thrombocytosis
- Highest risk period for coronary artery aneurysm

Convalescent Phase (Weeks 6–8)

- Clinical recovery
- ESR gradually normalizes
- Beau lines may appear

6. Diagnostic Approach

Child with fever ≥5 days → No obvious source of infection → Look for the 5 principal features → ≥4 criteria? If Yes → Kawasaki Disease. If No → Consider Incomplete KD → ESR + CRP elevated? → Perform Echocardiography → Coronary abnormalities present? → Treat as Kawasaki Disease

7. Investigations

Initial Laboratory Tests

- CBC
- ESR
- CRP
- Liver function tests
- Serum albumin
- Electrolytes
- Urinalysis (sterile pyuria)

ECG

Evaluate for:

- Arrhythmia
- Myocarditis
- Conduction abnormalities

Cardiac Evaluation — Echocardiography (Essential)

Perform:

- At diagnosis
- 1–2 weeks
- 4–6 weeks

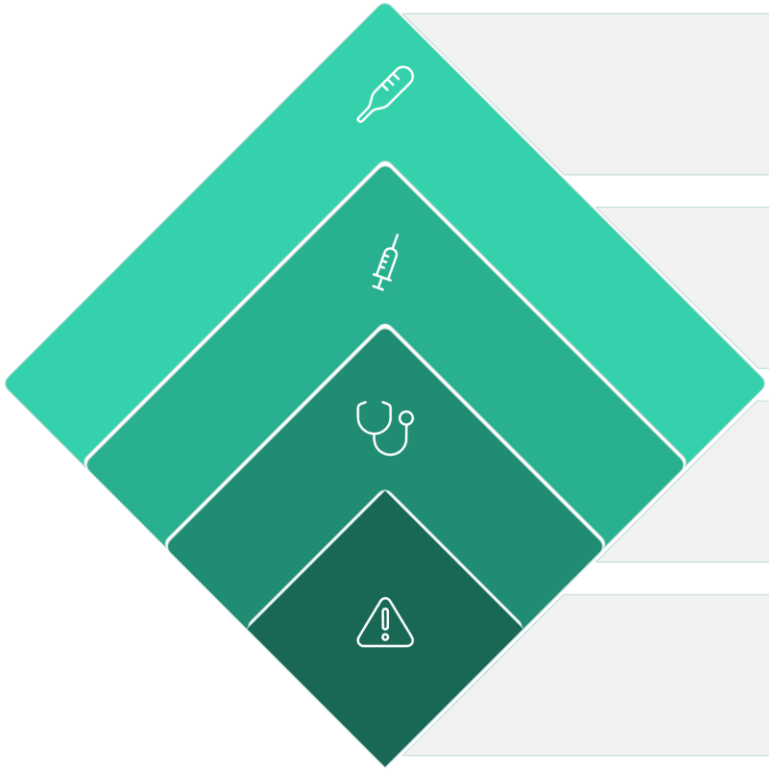
Evaluate:

- Coronary artery dilatation/aneurysm
- Ventricular function
- Pericardial effusion
- Valvular regurgitation

8. Differential Diagnosis

Disease	Distinguishing Features
Scarlet fever	Exudative pharyngitis, positive streptococcal test
Adenovirus	Purulent conjunctivitis
Measles	Cough, coryza, Koplik spots
Toxic shock syndrome	Hypotension, severe toxicity
MIS-C	Shock, GI symptoms, COVID-19 exposure
Systemic JIA	Daily spiking fever, arthritis

9. Management Algorithm



Reassess Fever

If afebrile 48–72h → adjust therapy

Initial Therapy

IVIG 2 g/kg + high-dose aspirin

Baseline Evaluation

Labs and echocardiography

Suspect KD

Do not delay treatment

Suspected Kawasaki Disease → Do NOT delay treatment → Obtain baseline laboratory tests → Perform Echocardiography → IVIG 2 g/kg (single infusion) + High-dose Aspirin → Afebrile for 48–72 hours? YES → Switch to Low-dose Aspirin → Repeat Echocardiography (1–2 weeks and 4–6 weeks). NO → Repeat IVIG → Consider Corticosteroids or Infliximab → Pediatric Cardiology Consultation

10. Treatment

Intravenous Immunoglobulin (IVIG)

- 2 g/kg IV
- Single infusion over 10–12 hours
- Ideally within the first 10 days of illness

Aspirin — Acute Phase

- 30–50 mg/kg/day (or 80–100 mg/kg/day, according to local protocol)
- Divided every 6 hours

After Fever Resolves

- 3–5 mg/kg/day once daily
- Continue until 6–8 weeks
- Continue longer if coronary artery abnormalities persist

IVIG-Resistant Kawasaki Disease

Persistent fever >36 hours after IVIG:

- Repeat IVIG
- Corticosteroids
- Infliximab (selected patients)

11. Complications

Cardiac

- Coronary artery aneurysm ⭐ (most important)
- Myocarditis
- Pericarditis
- Valvulitis
- Myocardial infarction (rare)

Other

- Arthritis
- Gallbladder hydrops
- Hearing loss (rare)

12. Follow-Up

All patients require:

- Pediatric cardiology follow-up
- Repeat echocardiography
- Long-term follow-up if coronary abnormalities are present

13. Red Flags

Urgent pediatric cardiology consultation if:

- **Shock**
- **Heart failure**
- **Persistent fever after IVIG**
- **Coronary artery aneurysm**
- **Arrhythmia**
- **Severe ventricular dysfunction**

14. Clinical Pearls

✓ Think of KD in any child with fever ≥5 days without an identifiable source.

✓ Diagnosis is clinical.

✓ Do not wait for laboratory confirmation before treatment.

✓ Echocardiography is mandatory in all suspected cases.

✓ IVIG within the first 10 days dramatically reduces coronary artery aneurysms.

✓ The subacute phase carries the highest risk for coronary artery aneurysm.

✓ Incomplete KD is more common in young infants and has a higher risk of coronary complications.

15. One-Page Take-Home Message

Remember: FEVER + 5

Fever ≥5 days PLUS 4 of the following 5 signs:

- **Bilateral conjunctivitis**
- **Oral changes**
- **Polymorphous rash**
- **Extremity changes**
- **Cervical lymphadenopathy**

Essential Investigations

- CBC
- ESR
- CRP
- Liver function tests
- Urinalysis
- Echocardiography

Treatment

1. IVIG 2 g/kg IV (single dose)
2. High-dose aspirin
3. Low-dose aspirin after fever resolves
4. Repeat echocardiography at 1–2 weeks and 4–6 weeks

Must Remember

- Most important complication: Coronary artery aneurysm
- Best investigation: Echocardiography
- Best treatment: IVIG within the first 10 days of illness
- Never delay treatment while waiting for laboratory results

Exam Pearls

- Cause: Medium-vessel vasculitis
- Most affected age: <5 years
- Diagnosis: Fever ≥5 days + ≥4 of 5 clinical criteria
- Gold-standard cardiac investigation: Echocardiography
- Treatment of choice: IVIG + Aspirin
- Most serious complication: Coronary artery aneurysm