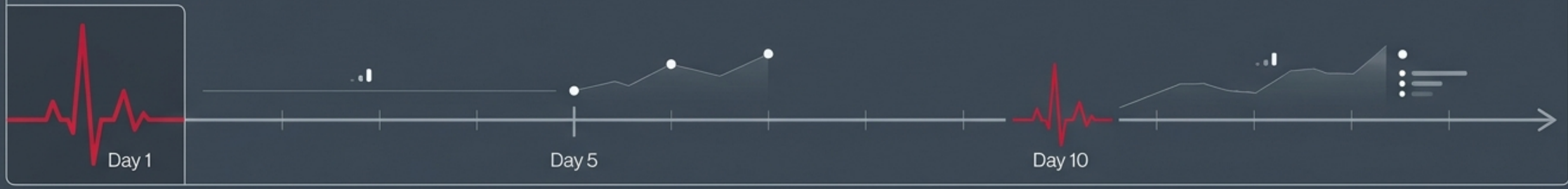
KDR

AUSTRALIAN CONTEXT

Kawasaki Disease & The 10-Day Intervention Window

A rapid clinical reference for diagnosis, risk stratification, and timely management in the Australian context.



Epidemiology

8-10 per 100,000 annual incidence in Australian children under 5.

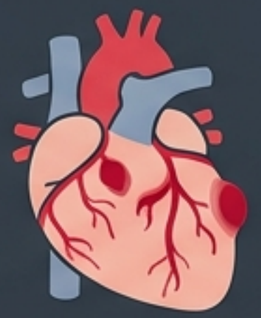
- 80% of cases are <5 years old.
- Peak incidence is 18-24 months.
- Boys affected 1.5x more than girls.
- Peaks in winter/spring.

The Primary Threat

Coronary Artery Aneurysms (CAA)

Occurs in ~25% of untreated children.

Risk is highest in infants <1 year and males.

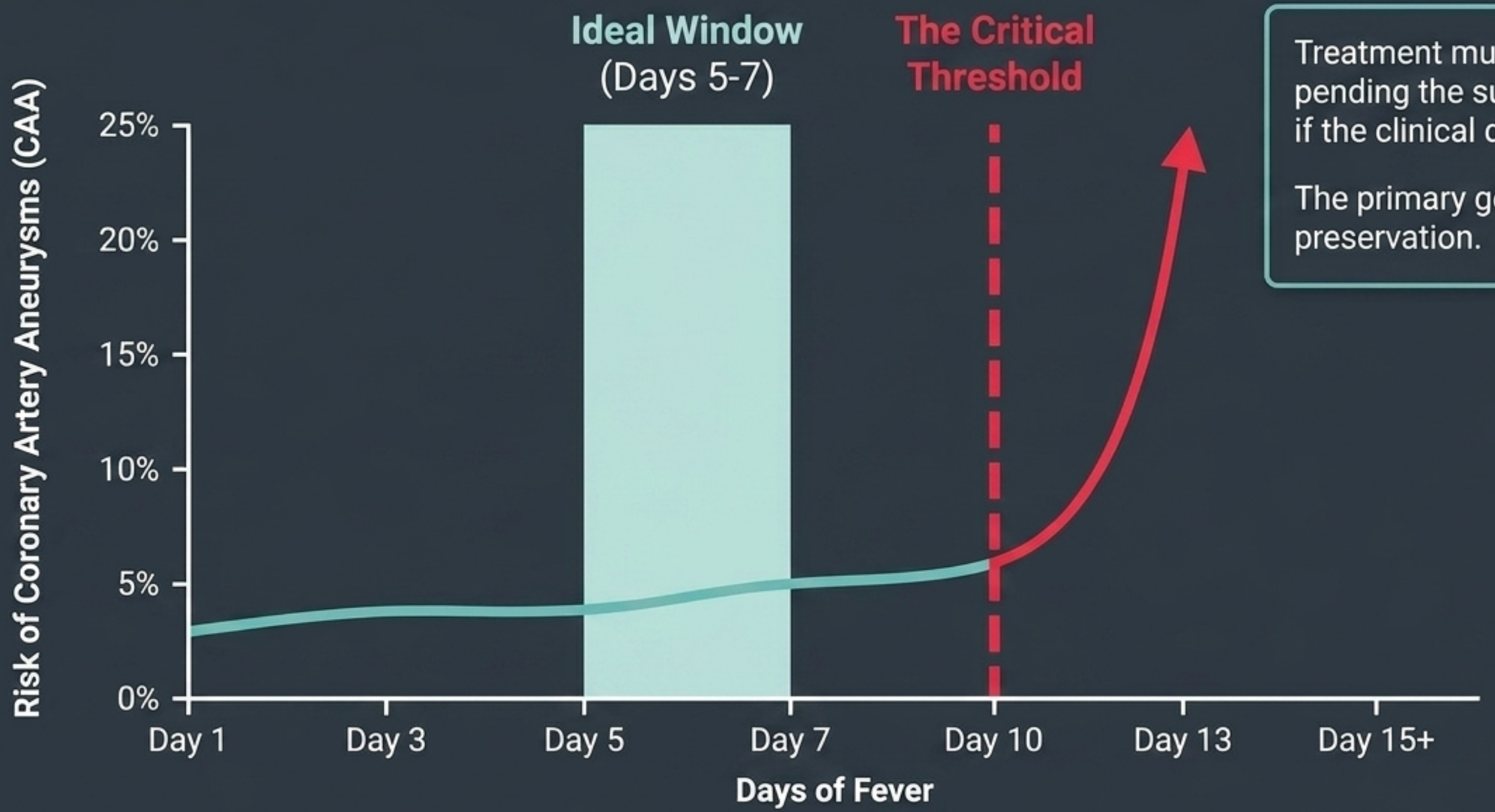


The Diagnostic Pitfall

KD is the disease that mimics many others.

Delayed diagnosis is the single greatest driver of irreversible coronary artery complications.

Day 1-4: Fever Onset



Treatment must not be delayed pending the subsidence of fever if the clinical diagnosis is clear.

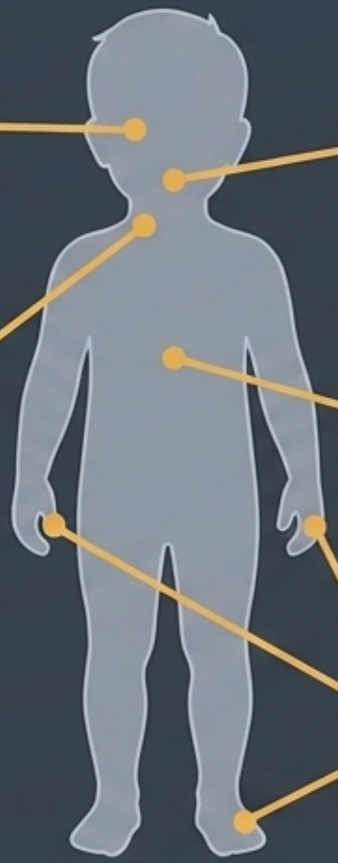
The primary goal is vascular preservation.

Days 5-10

Requirement: Persistent Fever ≥5 Days

1. **Eyes:** Bilateral non-exudative conjunctival injection (bulbar, limbic sparing, painless).

2. **Mouth:** Changes of oral mucous membranes (erythema, cracked lips, strawberry tongue).



3. **Neck:** Cervical lymphadenopathy (≥1.5 cm, usually unilateral, least common feature at 50-75%).

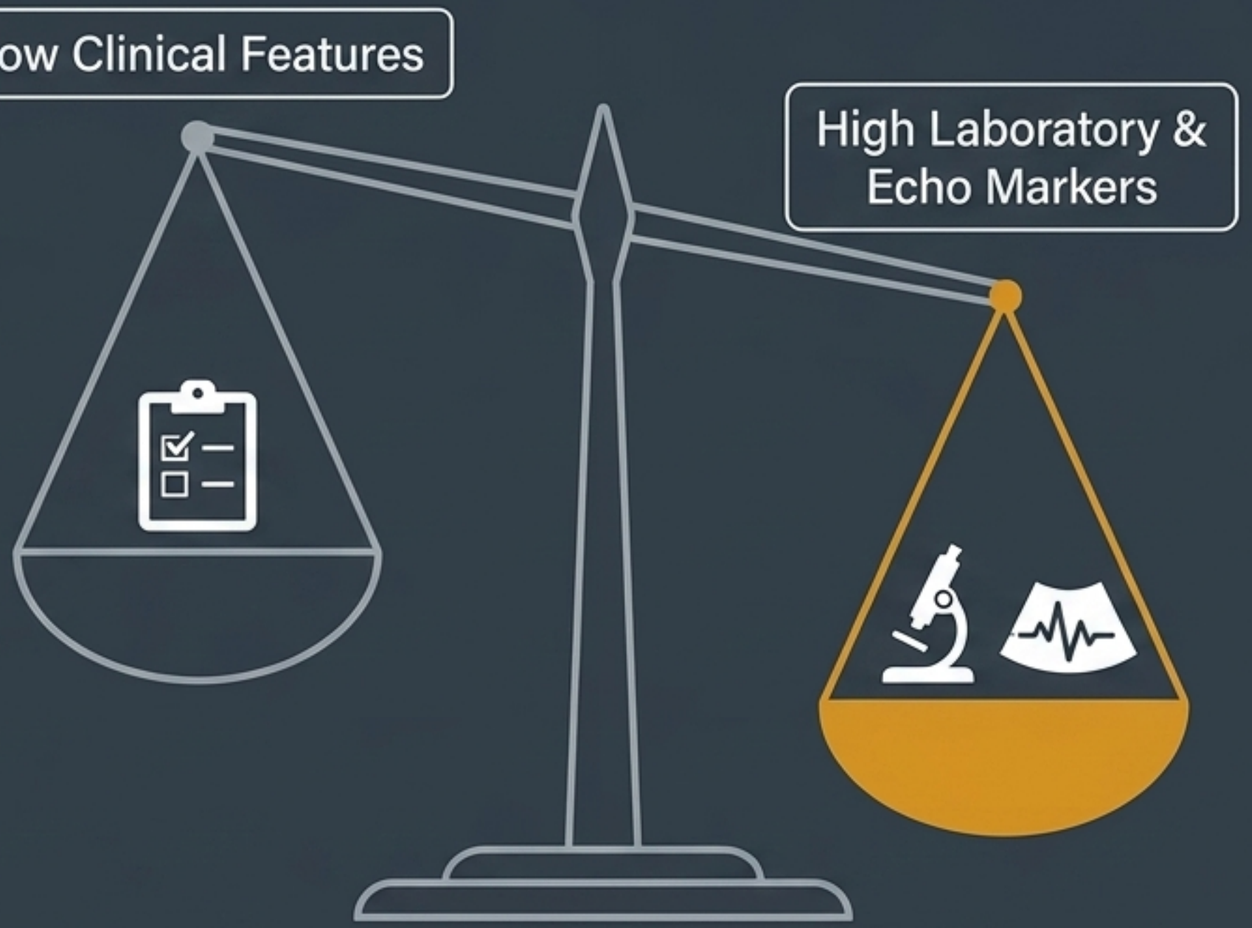
4. **Trunk:** Polymorphous rash (maculopapular, diffuse erythroderma, no vesicles/crusts).

5. **Extremities:** Acute erythema/oedema of hands and feet. (Note: periungual peeling is subacute weeks 2-3).

Diagnosis is clinical. 4 of 5 features + 5 days of fever = Classic KD.
(Fever can be <5 days if features are present and other causes excluded).

Incomplete Kawasaki Disease

Applies to persistent unexplained fever with <4 principal features.



WARNING: Infants <6 months are at the highest risk for CAA but frequently present with incomplete features. Incomplete does not mean milder—it represents the highest risk profile for giant aneurysms due to delayed recognition. Atypical features (shock, meningitis, uveitis) mandate a low threshold for treatment.

1. Clinical Suspicion

- Fever ≥4 days + any principal feature
  OR
- Unexplained fever ≥7 days in an infant

2. Laboratory Gateways

Primary Gate: CRP ≥30 mg/L and/or ESR ≥40 mm/hr.

Supplementary Gate
(Require ≥3 of 6 if primary met):
Albumin ≤30 g/L
Anaemia
ALT >45 U/L
Platelets (after day 7) >450 $x10^9$/L
WCC ≥15 $x10^9$/L
Urine WBC ≥10/HPF

3. Echocardiography & Action

- If ≥3 supplementary labs met OR CRP/ESR persistently elevated -> Treat as KD

Critical Note: Echo is for diagnostic support and baseline risk stratification; it is NOT a gatekeeper for treatment if clinical suspicion is high.

Normal
Z-score < 2

Smooth, normal lumen.

Dilation
Z-score 2.0 to < 2.5

Slight widening, maintaining shape.

Small Aneurysm
Z-score ≥2.5 to < 5

Focal or diffuse ballooning. Potential for stenosis.

Giant Aneurysm
Z-score ≥10 (or >8 mm)

Massive irregular ballooning. Turbulent flow.

Echocardiography measures coronary arteries against body surface area to generate a z-score (standard deviations from the mean). A Z-score ≥10 represents the **highest risk** of **thrombosis, myocardial infarction**, and **death**.

Baseline Echo

Intravenous Immunoglobulin (IVIG)

Dose: 2 g/kg as a single infusion over 10-12 hours. (Weight-based).

Timing: Must administer before Day 10 of illness.

Precautions: Monitor for infusion reactions. Defer live vaccines (MMR/Varicella) for 11 months.

Status: PBS Authority Required.

Aspirin (Acetylsalicylic Acid)

Acute Phase: 80-100 mg/kg/day orally (divided in 4 doses) until afebrile for 48-72 hours.

Subacute Phase: 3-5 mg/kg/day orally (once daily) until 6-8 week follow-up echo shows no aneurysm.

Australian Context Warning: High-dose aspirin remains standard practice in AU guidelines. Counsel parents on Reye's syndrome risk; avoid OTC NSAIDs/aspirin during treatment.

The Refractory Patient

Persistent or recrudescent fever 36 hours to 7 days after completion of the first IVIG infusion. Occurs in 10-20% of patients. High risk for CAA.

Path 1 (Most Common): Second Dose IVIG

2 g/kg IV single infusion.
~70% response rate.

Path 2 (Targeted Biologic): Infliximab

5 mg/kg IV single infusion. (TNF-α inhibitor, requires specialist PBS authority).

Path 3 (Severe/Rescue): Pulse Methylprednisolone

30 mg/kg/day IV (max 1g) for 1-3 days, followed by oral taper. Modern data supports safety despite historical aneurysm concerns.

Post-Treatment Window

The Infant (<6 months)

Pattern: Frequently presents with incomplete features. Diagnosis is often delayed. Highest biological risk for **giant aneurysms**.

The Refractory Patient

Pattern: Fails initial IVIG. Indicated by high baseline inflammatory markers (CRP/ESR), low albumin, low haemoglobin, and low sodium at presentation.

Prolonged inflammation damages vessels.

The Remote Indigenous Child

Pattern: Incidence is equal or higher, but **outcomes** are threatened by geographic barriers. **Delayed presentation, lack of immediate point-of-care echocardiography**, and challenges with continuity of cardiac follow-up.

Care requires culturally safe coordination with ACCHOs and retrieval services.

Risk Stratification

	Classic Kawasaki Disease	Typical MIS-C
Epidemiology	Peak <5 years, Asian ancestry	School-age median ~8 years, all ethnicities
Serology	Not linked to COVID	SARS-CoV-2 PCR or antibody positive in ~80-90%
Presenting Features	Classic mucocutaneous	Severe abdominal pain, vomiting, diarrhea, shock
Cardiac Involvement	Coronary artery aneurysms / medium vessels	Myocarditis, ventricular dysfunction, valvulitis
Laboratory Profile	High CRP/ESR, later thrombocytosis	Very high CRP/Ferritin/D-dimer, lymphopenia, thrombocytopenia
Treatment Response	IVIG alone is effective first-line	IVIG alone insufficient, requires systemic corticosteroids ± biologics

Differential Considerations

Level I (No Involvement)	Z-score <2.0	Normal lumen	Discharge from cardiology after 6-8 week echo. Healthy lifestyle counselling.
Level II (Dilation Only)	Z-score 2.0 to <2.5	Slight widening	Low-dose aspirin until regression confirmed. Repeat echo at 1 year.
Level III (Small Aneurysm)	Z-score ≥2.5 to <5	Focal or diffuse ballooning	Low-dose aspirin long-term. Annual cardiology review. Stress testing at transition to adult care.
Level IV (Medium/Giant Aneurysm)	Z-score ≥5	Massive ballooning	Low-dose aspirin + anticoagulation (Warfarin/DOAC). 6-12 monthly cardiology Lifelong follow-up and structured transition to adult congenital heart disease specialist. Pregnancy counselling required for females.

Weeks to Years

References

American Heart Association (AHA) Scientific Statement (McCrindle et al., 2017).

Australian Institute of Health and Welfare (AIHW): Kawasaki disease in Australia (2021).

Children's Health Queensland Clinical Practice Guideline (2022).

National COVID-19 Clinical Evidence Taskforce: MIS-C (2022).

RHDAustralia Guideline (3rd edition, 2020).

Data source: Med2Date Clinical Guidelines (Rheumatology/Paediatric).