Ashford and St. Peter's Hospitals
NHS Foundation Trust

Investigation and Management of Kawasaki Disease

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Guideline History		
Date	Comments	Approved By
November 2014	New Guideline	Paediatric Guideline Group
March 2018	Whole document review – no changes	Paediatric Guideline Group
June 2022	Whole document review - Revision of guidelines in keeping with AHA and Share guidelines	Paediatric Guideline Group
November 2022	Rewording of aspirin management	Dr Claire Mitchell
January 2026	Minor changes in line with network advise	Paediatric Guidelines Group

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1.

Investigation and management of Kawasaki Disease

Introduction

Kawasaki Disease (KD) is an acute vasculitic syndrome of uncertain aetiology, predominantly affecting children under 5 years of age. It is the second most common vasculitis of childhood (after HSP) and is the most common cause of acquired heart disease in children i developed countries. Up to 25% of untreated patients may develop coronary artery aneurysms, declining to approximately 4% with early intervention.
Where the inflammatory aetiology is unclear, please refer to pyrexia of unknown origin guideline, consider PIMS-TS and Macrophage Activation Syndrome as potential differential diagnoses.

Classic clinical criteria – complete KD

Fever persisting for >5 days * AND at least 4 of the following	
Conjunctival injection	Often bilateral, non-purulent conjunctivitis; bulbar; limbic sparing typical
Oro-mucosal changes	Cracked red lips, strawberry tongue, diffuse erythema of the oropharynx,
Rash	Polymorphous widespread rash (maculopapular, scarlatiniform, urticarial) but not vesicular or petechial predominant.
Affected hands and feet	Acute phase – oedema and/or erythema; Convalescent phase – desquamation
Lymphadenopathy	Cervical, Usually unilateral; at least one lymph node ≥ 1.5 cm in diameter

*If all criteria are met prior to 5 days duration of fever do not delay treatment as earlier recognition and management is associated with better outcomes.

Other associated features may include marked irritability, induration and erythema around the BCG scar, arthritis, uveitis, gastroenteritis and dysuria

Recommended Investigations

Blood tests:

- FBC, Film, UEs, LFTs
- ESR*, CRP (*ESR is only useful prior to IVIG)
- Ferritin, LDH, CK - not routine unless assessing for PIMS-TS or MAS •

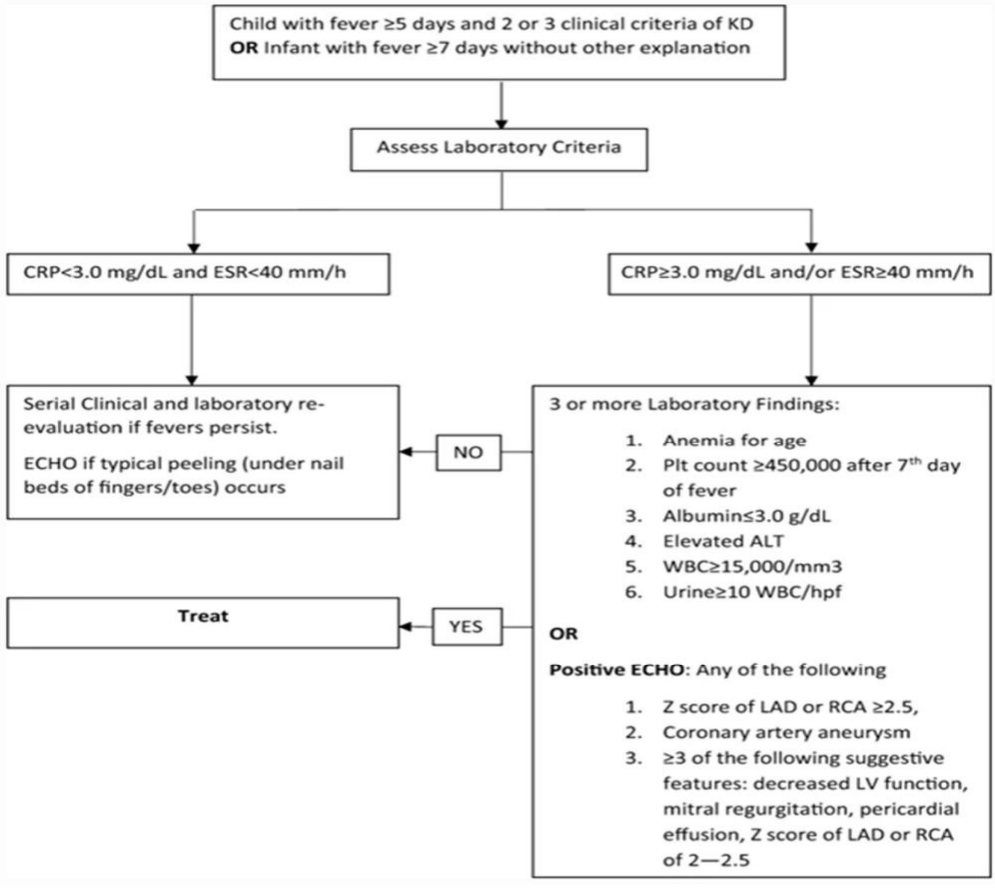
- Blood Culture
- ASO titre and AntiDNAse B
- Viral titres (enterovirus, adenovirus, CMV, EBV, SARS-CoV2 IgG) • Autoantibody screen (ANA)
- Consider differential diagnosis of PIMS-TS

Further investigations:

- Nasal and throat swabs – bacterial and viral
- Urine microscopy
- CXR
- Abdominal US – for evidence of organomegaly and systemic involvement
- 12 lead ECG
- Consider Echocardiogram at time of diagnosis or if incomplete criteria met. Discuss with Dr Groves or technician depending on age.

Incomplete KD

Initial presentation may not meet all the above criteria, but these patients are still at risk of complications from the disease and require treatment. See AHA algorithm for incomplete KD.



Management of Kawasaki Disease

If further advice is required and diagnosis is unclear, discussion with the Paediatric infectious disease team at St Georges is recommended. If Diagnosis is clear but there are worrying features/poor response to treatment discuss with Royal Brompton Paediatric Cardiology team.

First line treatment:

A. Immunoglobulin: 2 g/kg IV single dose infusion based on ideal body weight usually over 10 - 12 hours

B. Aspirin: Acute Phase: 30-50 mg/kg/day (in 4 divided doses)

After defervescence: 3-5 mg /kg/day for 6 weeks minimum for all patients. If initial echocardiogram abnormal then continue aspirin until echocardiogram normalises and inflammatory markers normalise . If coronary artery involvement persists get advice as per Cardiology

(See SHARE guidelines)

C. Consider Corticosteroids in high risk patients in discussion with tertiary unit (Discussion with RBH)

(Recommended likely treatment– 2nd IVIG dose, IV methylprednisolone 10mg/kg (max 1g) OD for 3 days then weaning course of oral prednisolone OR infliximab)

Steroids should be given upfront with first IVIG (in discussion with tertiary unit) in:

- age <12 months
- features of shock or macrophage activation syndrome
- high CRP (>100)
- early coronary changes
- IVIG resistance – persistant or rescrudescent fever at least 36 hours after completion of IVIG

Assessing response to treatment

Defervescence + improving clinical features + normalising CRP.

If inflammation persists second line treatment must be considered (as well as differential diagnosis). Discuss with PID at St Georges and Cardiology at Royal Brompton Hospital. ECG in Kawasaki Disease

- At presentation
- Prior to discharge
- With severe episodes of pain or discomfort

Echocardiogram

Must be done within 2 weeks for typical KD that has responded to treatment. Repeat echo more urgently if no response to IVIG or signs of persistent inflammation (fever/ CRP/ clinical concern). Dates below correlate to follow up intervals with Dr Groves.

Initial echo – showing NO evidence of coronary involvement,

- Repeat at 6-8 weeks
- Repeat at 6 months
- Annually until old enough to do exercise ECG

Initial echo – showing EVIDENCE of coronary involvement (Aneurysms)

- Referral to tertiary centre (Royal Brompton Hospital Cardiology)

After initial echo repeat at 10-14 days if persistently high CRP, persistent fever or high risk features

Management of cardiac involvement in Kawasaki Disease

Risk factors for coronary artery involvement:

- White cell count >30
- ESR >100
- Male
- <1 year old
- Fever >16 days
- IVIG resistance

Echocardiogram:

- LCA, RCA proximal and distal segments
- LV function
- Pericardial effusion
- Valvar regurgitation

For patients with aneurysms, following tertiary discussion (RBH), consider treatment with LMW heparin acutely and longer term warfarin depending on age, size of aneurysm and cardiology recommendations.

Further consideration:

- After IVIG live vaccines should be delayed by 3 months
- The live intranasal flu vaccine is contraindicated for children on aspirin
- In children who have not had chicken pox or the vaccine there is a theoretical risk of developing Reye's syndrome whilst on aspirin if they are exposed to chicken pox. Advise families to present to local hospital if CYP develops persistent vomiting/ lethargy/ seizures or change in behaviour.

- Advised to represent if fever or rash returns (risk of relapse following treatment)
- Peeling of fingers or toes around week 2–3 is expected and not a sign of relapse.

Supporting References

- *N. de Graeff, N. Groot, D. Eleftheriou et al. European consensus-based recommendations for the diagnosis and treatment of Kawasaki Disease – the SHARE initiative. Rheumatology. 2019;58:672– 682.*
- *Diagnosis, treatment and long-term management of Kawasaki Disease: A scientific statement for health professionals from the American Heart Association (AHA).*
- *S. Benseler, B. McCrindle, E. Silverman et al. Infections and Kawasaki Disease: Implications for coronary artery outcome. Pediatrics. 2005;116(6):e760–e766.*
- *NICE Guideline NG143. Fever in under 5s: assessment and initial management. National Institute for Health and Care Excellence (NICE), 2019 (updated).*
- *NICE Evidence Review. Signs and symptoms predicting Kawasaki Disease in children with fever ≥5 days. National Institute for Health and Care Excellence (NICE).*
- *PIER Network (Paediatric Innovation, Education & Research Network). Kawasaki Disease: Diagnosis, Management, Follow-up & Referral Guidance.*
- *London Kawasaki Disease Research Group. Kawasaki Disease: an evidence-based approach to diagnosis, treatment and proposals for future research. Archives of Disease in Childhood. 2002.*

Supporting relevant trust guidelines

- *Pyrexia of Unknown origin*
- *PIMS-TS*
- *Paediatric Echocardiography Referral & Follow-up Pathway – Trust Guideline*
- *Paediatric Sepsis Recognition & Management Guideline – Trust Guideline*
- *PIER Network (Paediatric Innovation, Education & Research Network) – Kawasaki Disease: Diagnosis, Management, Follow-up & Referral*
- *Great Ormond Street Hospital (GOSH) – Kawasaki Disease Clinical Pathway*
- *Evelina London (GSTT) – Kawasaki Disease: Assessment and Early Management Guideline*
- *Royal Brompton Hospital / RBH Paediatric Cardiology – Coronary Artery Involvement in Kawasaki Disease: Anticoagulation & Follow-up Protocol*

2. Guideline Governance

a. Scope

This guideline is relevant to all staff caring for all children from 0-18 years old across the emergency department, inpatient ward and outpatient department. b. Purpose

i. This guidelines aims to facilitate a common approach to the management of children. At times deviation from the guideline may be necessary, this should be documented and is the responsibility of the attending consultant.

ii. This guideline is subject to regular review to ensure ongoing evidence based practice.

c. Duties and Responsibilities

All healthcare professionals responsible for the care of all children 0-18years should be aware of practice according to this guideline.

d. Approval and Ratification

This guideline will be approved and ratified by the Paediatric Guidelines Group.

e. Dissemination and Implementation

i. This guideline will be uploaded to the trust intranet 'Paediatric Guidelines' page and thus available for common use.

ii. This guideline will be shared as part of ongoing education within the Paediatric Department for both medical and nursing staff.

iii. All members of staff are invited to attend and give comments on the guideline as part of the ratification process.

f. Review and Revision Arrangements

a. This policy will be reviewed on a 3 yearly basis by the appropriate persons.

b. If new information comes to light prior to the review date, an earlier review will be prompted.

c. Amendments to the document shall be clearly marked on the document control sheet and the updated version uploaded to the intranet. Minor amendments will be ratified through the Paediatric Guidelines Group. A minor amendment would consist of no major change in process, and includes but is not limited to, amendments to documents within the appendices.

g. Equality Impact Assessment

Background Who was involved in the Equality Impact Assessment
Dr Alexandra Jones and Dr Alison Groves
Methodology • A brief account of how the likely effects of the policy was assessed (to include race and ethnic origin, disability, gender, culture, religion or belief, sexual orientation, age) The data sources and any other information used • The consultation that was carried out (who, why and how?)
All groups of staff and patients were taken into consideration and there is no bias towards or against any particular group.
Key Findings • Describe the results of the assessment • Identify if there is adverse or a potentially adverse impacts for any equalities groups
There is no evidence of discrimination.
Conclusion Provide a summary of the overall conclusions
There is no evidence of discrimination.

Recommendations
• State recommended changes to the proposed policy as a result of the impact assessment • Where it has not been possible to amend the policy, provide the detail of any actions that have been identified • Describe the plans for reviewing the assessment
This guideline is appropriate for use.

h. Document Checklist

To be completed (electronically) and attached to any document which guides practice when submitted to the appropriate committee for approval or ratification.

Title of the document: Investigation and management of Kawasaki Disease

Policy (document) Author: Dr Alexandra Jones

Executive Director: N/A

		Yes/No/ Unsure/NA	Comments
1.	Title		
	Is the title clear and unambiguous?	Y	
	Is it clear whether the document is a guideline, policy, protocol or standard?	Y	
2.	Scope/Purpose		
	Is the target population clear and unambiguous?	Y	
	Is the purpose of the document clear?	Y	
	Are the intended outcomes described?	Y	

	Are the statements clear and unambiguous?	Y	
3.	Development Process		
	Is there evidence of engagement with stakeholders and users?	Y	
	Who was engaged in a review of the document (list committees/ individuals)?	Y	Authors and guideline committee with backing from tertiary services
	Has the policy template been followed (i.e. is the format correct)?	Y	
4.	Evidence Base		

	Is the type of evidence to support the document identified explicitly?	Y	
		Yes/No/ Unsure/NA	Comments
	Are local/organisational supporting documents referenced?	Y	
5.	Approval		
	Does the document identify which committee/group will approve/ratify it?	Y	Paediatric guideline committee
	If appropriate, have the joint human resources/staff side committee (or equivalent) approved the document?	Y	
6.	Dissemination and Implementation		

	Is there an outline/plan to identify how this will be done?	Y	
	Does the plan include the necessary training/support to ensure compliance?	Y	
7.	Process for Monitoring Compliance		
	Are there measurable standards or KPIs to support monitoring compliance of the document?	Y	
8.	Review Date		
	Is the review date identified and is this acceptable?	Y	3 years
9.	Overall Responsibility for the Document		
	Is it clear who will be responsible for coordinating the dissemination, implementation and review of the documentation?	Y	
10.	Equality Impact Assessment (EIA)		
	Has a suitable EIA been completed?	Y	

Committee Approval (Paediatric Guidelines Group)			
If the committee is happy to approve this document, please complete the section below, date it and return it to the Policy (document) Owner			
Name of Chair	Dr Alexandra Jones	Date	14/01/2026

Ratification by Management Executive (if appropriate)
If the Management Executive is happy to ratify this document, please complete the date of ratification below and advise the Policy (document) Owner
Date: n/a