

Kawasaki Disease Guidelines for Diagnosis & Management

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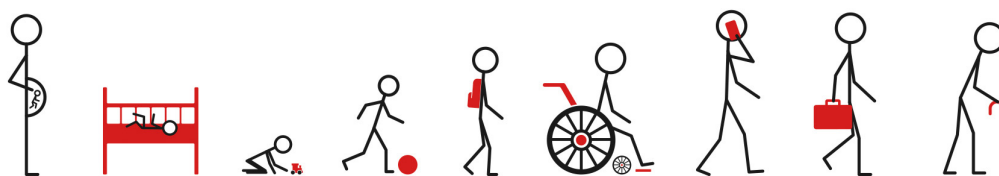


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Introduction

- Kawasaki disease (KD) is an acute systemic inflammatory vasculitis of medium sized arteries.
- Affects predominantly children below the age of five and is the commonest cause of acquired heart disease in childhood.
- Most concerning complication of KD is coronary artery aneurysm.
- Early recognition and early suppression of inflammation prevents morbidity and mortality related to coronary artery aneurysm.
- The aim of treatment is to switch off the inflammatory process, aiming for zero fever, zero CRP (or at least CRP halving every 24 hours) within 48 hours of starting treatment. Failure to achieve this target should prompt clinicians to consider the disease as refractory and consider additional rescue treatment.

Epidemiology

- Affects around 300 children per year in the UK and Ireland.
- Second commonest vasculitis of childhood after IgA Vasculitis (Henoch-Schonlein Purpura).
- More common in boys, with an ethnic bias towards Asian children.
- Peak incidence in winter and spring months.
- Approximately 85% of children with KD are younger than 5 years of age, with peak age incidence at 18-24 months but can occur in children even under 6 months.

Diagnosis

- There are no diagnostic tests for KD.
- Diagnosis rests on a combination of clinical criteria and laboratory findings.
- Children may present with the full complement of described features (complete Kawasaki disease) or only some features (incomplete Kawasaki disease).
- Children with particular “high risk” features at presentation are more likely to develop coronary artery aneurysms (CAA’s).
- Rising rates of coronary artery aneurysms in the UK and worldwide are felt to reflect delayed recognition and therefore delayed treatment.

Diagnostic criteria

- A.** Fever – duration of 5 days or more (typically sudden onset, swinging and including high grade fevers)

PLUS

B. Four out of five of the following:

1. Conjunctivitis – bilateral, bulbar, non-suppurative
2. Lymphadenopathy – cervical, often >1.5cm, often unilateral
3. Rash – polymorphous, no vesicles or crusts
4. Changes in lips or oral mucosa – red, cracked lips, ‘strawberry’ tongue, or diffuse erythema of oropharynx.
5. Changes of extremities – initial stage: erythema and oedema of palms and soles. Convalescent stage: peeling of skin from fingertips.

Further diagnostic considerations

- Diagnosis of ‘incomplete KD’ cases depends on a high level of suspicion in children presenting with some of the KD features and evidence of systemic inflammation (e.g. elevated ESR, CRP or leukocytosis).
- Incomplete cases are more likely to be at outside the typical KD age range and more likely to develop CAAs.
- Diagnosis of KD should not be delayed, and treatment instituted even when the fever is less than 5 days old if:
 - > Four out of above five diagnostic criteria of KD are present before day 5 of fever.
 - > Coronary artery aneurysm or coronary dilatation are present.
 - > Evidence of persistent elevation of inflammatory marker with no other explanation in patients where there remains clinical suspicion of KD (see incomplete KD section and diagram 1, p9).
 - > Seek expert Cardiologist/Rheumatologist advice in such cases.
- Misery is an important sign which is nearly always present.

Other possible clinical findings in KD

	Moderately or Less Common	Uncommon or Rare
Cardiovascular	Myocarditis, pericarditis, valvular regurgitation	
	Coronary artery abnormalities	
	Aneurysms of medium-sized non coronary arteries	
	Peripheral gangrene	
	Aortic root enlargement	
	Kawasaki Shock Syndrome	
Respiratory	Peri-bronchial and interstitial infiltrates on chest x-ray	
	Pulmonary nodules	
	Pneumonitis	

Musculoskeletal	Arthritis, arthralgia	
Gastrointestinal	Diarrhoea, vomiting, abdominal pain	Gallbladder hydrops
	Hepatitis	Gastrointestinal ischaemia
	Pancreatitis	Jaundice
Nervous system	Extreme irritability	Cranial Nerve Palsy
	Aseptic meningitis	Febrile Convulsions
		Encephalopathy
		Ataxia
		Sensorineural hearing loss
Renal or Genitourinary	Urethritis/meatitis, hydrocele	Tubular Interstitial Nephritis or non-inflammatory tubular disturbances
	Pyuria and or proteinuria	Renal Failure
Other	Desquamating rash in groin	Petechial rash
	Retropharyngeal inflammation	
	Anterior uveitis on slit lamp examination	
	Erythema and induration at BCG site *	

(Adopted from Newburger et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Statement for Health Professionals From the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. *Pediatrics* 2004;114(6):1708–33.)

* Erythema/induration at site of BCG scar is a very specific clinical finding and a useful sign to look for in uncertain/incomplete cases since it makes KD very likely. However, it has been shown that prevalence of this reaction in KD patients declines with time after the BCG vaccination and is much less prevalent beyond 12 months after the inoculation. Therefore, a non-reactive BCG scar should not reduce clinical suspicion of KD.

Differential diagnosis

- Streptococcal infection (Scarlet fever and Toxic Shock like Syndrome (TSS))
- Staphylococcal infection (TSS or Scalded Skin syndrome)
- Paediatric Inflammatory Multisystem Syndrome (PIMS-TS) (low worldwide incidence in 2024)
- Viral infection
 - > Measles
 - > Rubella
 - > Roseola infantum
 - > EBV infection
 - > Influenza A & B
 - > Adenovirus
- Mycoplasma Pneumonia infection
- Stevens-Johnson Syndrome
- Systemic onset idiopathic juvenile arthritis

- Haemophagocytic lymphocytic histiocytosis (HLH) of other cause
- Diagnostic pitfalls include mistaking
 - > Rash and mucosal changes for an antibiotic reaction
 - > Sterile pyuria for partially treated Urinary Tract Infections
 - > CSF pleocytosis for viral meningitis

*Kawasaki Disease is **less** likely in the presence of*

- exudative tonsillitis
- purulent conjunctivitis
- discrete intraoral lesions
- bullous or vesicular rash
- splenomegaly
- generalized lymphadenopathy

It is important to note that evidence of bacterial infection may be present in patients with KD (for example, evidence of recent Streptococcal infection) and clinicians may need to treat infection alongside KD. Children may also present with KD and co-existing viral infection for example (but not limited to) rhinovirus or enterovirus.

Incomplete KD

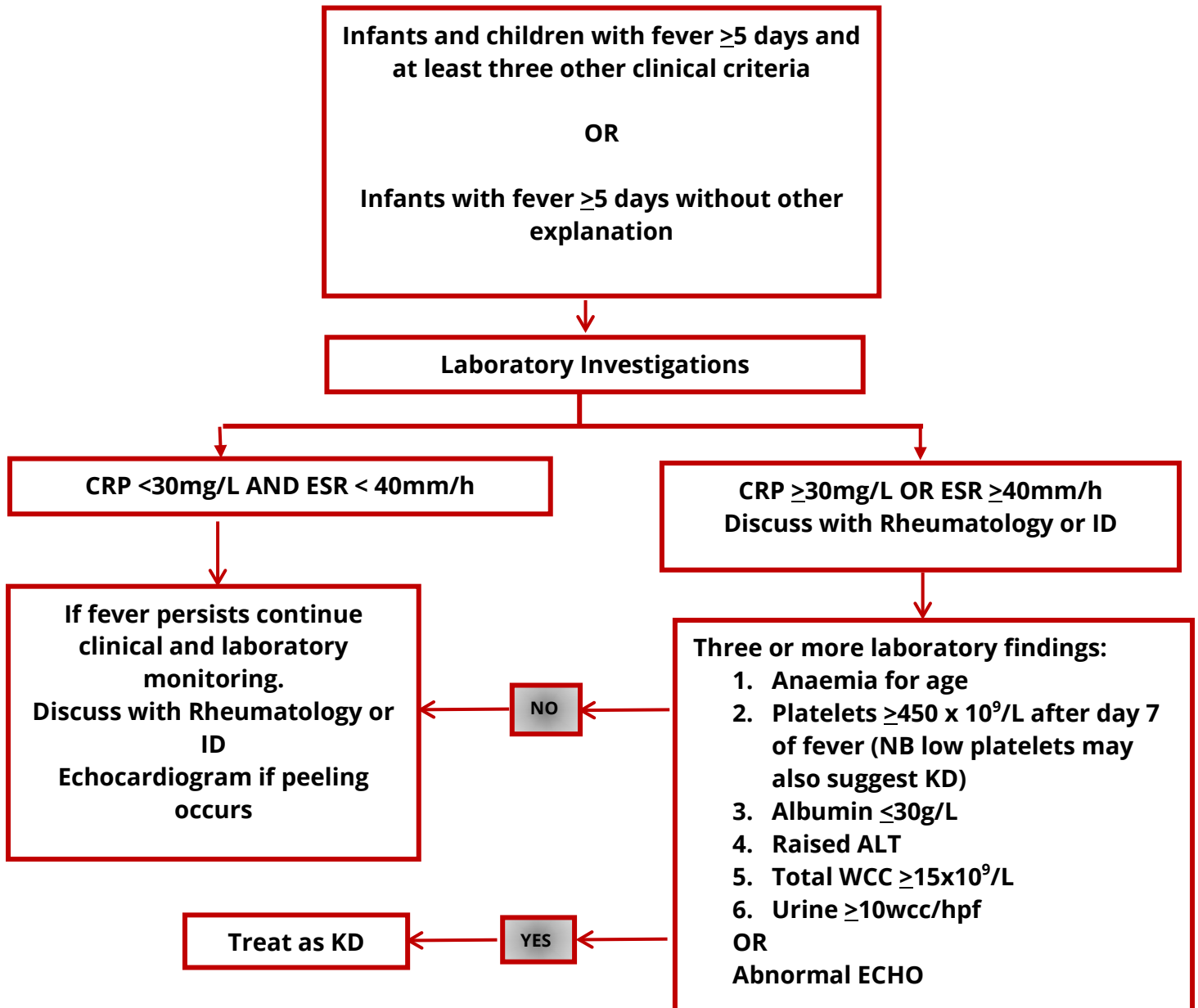
Diagnosis of incomplete KD requires a high degree of suspicion and should be considered in any child with a fever of ≥ 5 days. Many patients do not have all the classical clinical features or clinical features may be transient.

Supplemental criteria may support a diagnosis of incomplete Kawasaki disease in a child with fever < 5 days but meeting the other diagnostic criteria, OR fever ≥ 5 days but < 4 other diagnostic criteria.

Supplemental Criteria
Albumin ≤ 30 g/L
Raised ALT
White blood cells $> 15 \times 10^9/L$
Platelets $> 450 \times 10^9/L$ after day 7 of illness (or thrombocytopenia)
Anaemia for age
> 10 urine white blood cells/hpf

Management of suspected incomplete KD

Diagram 1



(From McCrindle et al, Diagnosis, treatment, and long-term management of Kawasaki disease: A scientific statement for health professionals from the American Heart Association. Circulation 2017;135(17):927-99, and De Graeff et al. European consensus-based recommendations for the diagnosis and treatment of Kawasaki disease-the SHARE initiative. Rheumatol (Oxford) 2019;58(4):672-82.)

Clinical course

The course of **untreated** KD can be described in three clinical phases

- > **Acute phase:** lasts 7 – 14 days, characterized by fever and inflammatory changes.
- > **Subacute phase:** typically lasts from approximately day 10 to day 25 after onset of illness. Fever, rash and lymphadenopathy resolve even without treatment, but irritability, anorexia and conjunctival injection can persist along with arthritis, arthralgia or myocardial dysfunction. Regardless of treatment desquamation of fingers and toes appear, and thrombocytosis is common.
- > **Convalescent phase:** Begins when all clinical signs disappear and continues until acute phase reactants return to normal, usually six to ten weeks after onset.

Complications

- Coronary artery aneurysms (CAA) or coronary artery dilatation usually occurs within 6-8 weeks of onset of the illness, affecting around 20% of children with untreated KD. Patients who develop CAA are at risk of coronary artery thrombosis, myocardial infarction and sudden death. Angiography may be required to diagnose subsequent regression of CAA.
- Additional rare complications of KD include:
 - Macrophage activation syndrome/ secondary haemophagocytic lymphohistiocytosis (HLH) characterised by fever, hepatosplenomegaly, lymphadenopathy, rash, pancytopenia. If HLH is suspected, we would direct the clinician to the GIRFT 2024 best practice pathway for HLH.
 - Syndrome of inappropriate antidiuretic hormone secretion resulting in hyponatraemia.

Investigations

Investigations are aimed at identifying alternative diagnoses and assessment of complications.

- FBC, ESR, CRP
- U&Es, LFTs, Amylase
- Ferritin: Consider HLH if hyperinflammation, cytopenia and hyperferritinaemia
- Blood culture
- CSF MC&S if signs of meningitis
- Urine dipstick, MC&S
- Throat swab
- ECG, may see

- > ST segment depression
 - > T wave inversion
 - > Conduction disturbances Eg. Heart block
- Echocardiogram, may show
 - > Myocarditis
 - > Pericardial effusion
 - > Bright coronary arteries (experienced operators)
 - > Coronary artery aneurysm
- Consider ASOT level, resp. viral PCR panel, viral titres and mycoplasma serology
- Not all of the inflammatory markers may be abnormal at first presentation and repeat blood testing should be undertaken if there is diagnostic uncertainty.
- Thrombocytosis occurs towards the end of the 2nd week of the illness and therefore, may not be helpful in the early stages. Acute thrombocytopenia or low/normal platelet count may occur and may be associated with a poorer prognosis.
- LFTs may be deranged. Hypoalbuminaemia is common.
- Sterile pyuria and CSF pleocytosis (predominantly lymphocytes) representing aseptic meningitis also occur.

Timing of Echocardiogram

Echocardiogram should be performed

- At presentation.
- At 10-14 days of disease onset even if the initial echo was normal.
- Echo should be performed more frequently in those with aneurysm detected on initial echo and with ongoing active inflammation to monitor aneurysm size progression, or the development of thrombus formation. Discuss with Cardiology.
- At 6-8 weeks after disease onset.
- Then consider need for ongoing further echocardiography depending on presence/absence of coronary artery aneurysms (see “follow up” section and treatment flow chart)

Treatment

Acute phase

- 2 g/kg of **intravenous immunoglobulin (IVIG)** as a single IV infusion. The dose and rates of infusion are calculated on ideal body weight and are dependent on the brand of IVIG used. Refer to your local IVIG policy, NHSE IVIG commissioning criteria (<http://igd.mdsas.com/>) and appendix B of this document. You may also need to liaise with your local pharmacy department for calculation of the infusion rates.
- Patients with IVIG resistance or only partial response (ongoing fever and/or persistent inflammation) may be given a second dose of IVIG of 2g/kg. These

patients should be always discussed first with regional paediatric rheumatology service.

- **Aspirin** 30 – 50 mg/kg/day in four divided doses during the acute phase of illness (maximum *450mg/dose*) and then once fever and inflammation subsides, reduce the dose to an antiplatelet dose of 2-5 mg/kg/day (maximum 75mg/dose). It is recommended to reduce aspirin to the antiplatelet dose early if corticosteroids are commenced. In the rare case of neonatal Kawasaki disease, aspirin doses will differ for neonates (refer to BNFC).
- **Corticosteroids** are recommended in conjunction with IVIG in high risk and poorly responsive cases:
 - > patients resistant to IVIG treatment with ongoing fever, and /or persistent inflammation or clinical signs greater than 48 hours after receiving IVIG as a single dose of 2 g/kg.
 - > As first line treatment alongside IVIG in patients with features of most severe disease (and therefore the greatest likelihood of developing CAA):
 - a. Very young patients (less than 1 year old).
 - b. Those with markers of severe inflammation, including liver dysfunction, hypoalbuminaemia and anaemia.
 - c. Features of haemophagocytic lymphohistiocytosis (HLH).
 - d. Features of shock.
 - > Patients who already have evolving coronary and/or peripheral aneurysm with ongoing inflammation at presentation or extra-coronary manifestations such as mitral regurgitation or pericardial effusion.

Dose of corticosteroids

There is no consensus on the optimal regime of corticosteroid in KD. Based on the European consortium (SHARE guidelines) two treatment regimens are considered reasonable. Treating clinicians will need to determine the corticosteroid regimen for individual patients.

Regime 1: IV methylprednisolone 0.8 mg/kg BD for 5-7 days or until CRP normalizes; then convert to oral prednisolone 2 mg/kg/day (maximum per dose 40 mg)

Oral prednisolone should then be weaned slowly over next 2-3 weeks.

Or

Regime 2: IV methylprednisolone 10-30 mg/kg (maximum of 1 gram/day) once daily for three days; then convert to oral prednisolone 2 mg/kg/day (maximum per dose 40 mg) until day 7 or until CRP normalizes

Oral prednisolone should then be weaned slowly over next 2-3 weeks.

Individual cases should be discussed with paediatric rheumatology if there is uncertainty about the most appropriate corticosteroid regime.

Patients taking acute phase dose aspirin or starting corticosteroids should also be started on a proton pump inhibitor for gastric protection.

Biologics

Patients with KD resistant to treatment with IVIg, aspirin and corticosteroids may be considered for biologic treatment (see treatment flow sheet). Please discuss with the tertiary Rheumatology team and consider transfer to regional centre. Currently no biological treatments are commissioned by NHS England or Wales for the treatment of KD, so the decisions of whether to escalate to biological treatment and which to use may vary between tertiary centres.

Convalescent phase

- If coronary aneurysm has been identified, antiplatelet therapy, 2-5 mg/kg of once daily aspirin (max. 75mg/dose) should be continued at least until the aneurysm resolves.
- In the presence of medium or large aneurysm (>8 mm internal diameter; for infants Z score > 7 based on Montreal normative data) lifelong once daily aspirin at 2-5 mg/kg is recommended (maximum 75mg/dose).
- In addition, when giant aneurysms are identified, anticoagulation with warfarin and/or heparin is indicated and should be discussed with regional cardiology unit and regional paediatric haematology unit.
- Aspirin may be stopped at 6-8 weeks if the initial echocardiogram was normal and repeat echo at 6-8 weeks is normal.

Additional Considerations of Treatment

- Treatment **can** be commenced before 5 days of fever as outlined above in the diagnosis section, at the point a diagnosis of KD is made.
- Concomitant treatment of sepsis may also be indicated until sepsis is excluded.
- Treatment **should** be given if the presentation is greater than 10 days from fever onset if there are signs of persistent inflammation (which in untreated cases one would expect to find). Discuss all such delayed presentations with your local tertiary centre.
- Response to treatment with disease defervescence must be demonstrated by **both** resolution of fever and consistently falling CRP. Do not rely on resolution of fever alone as the sole determinant of therapeutic success. CRP should be repeated daily until it is normal (<10mg/L) or at least consistently halving each day and all clinical features have resolved. If fever resolves but CRP is static, please discuss with the tertiary rheumatology service as further treatment may be required.

Follow-Up

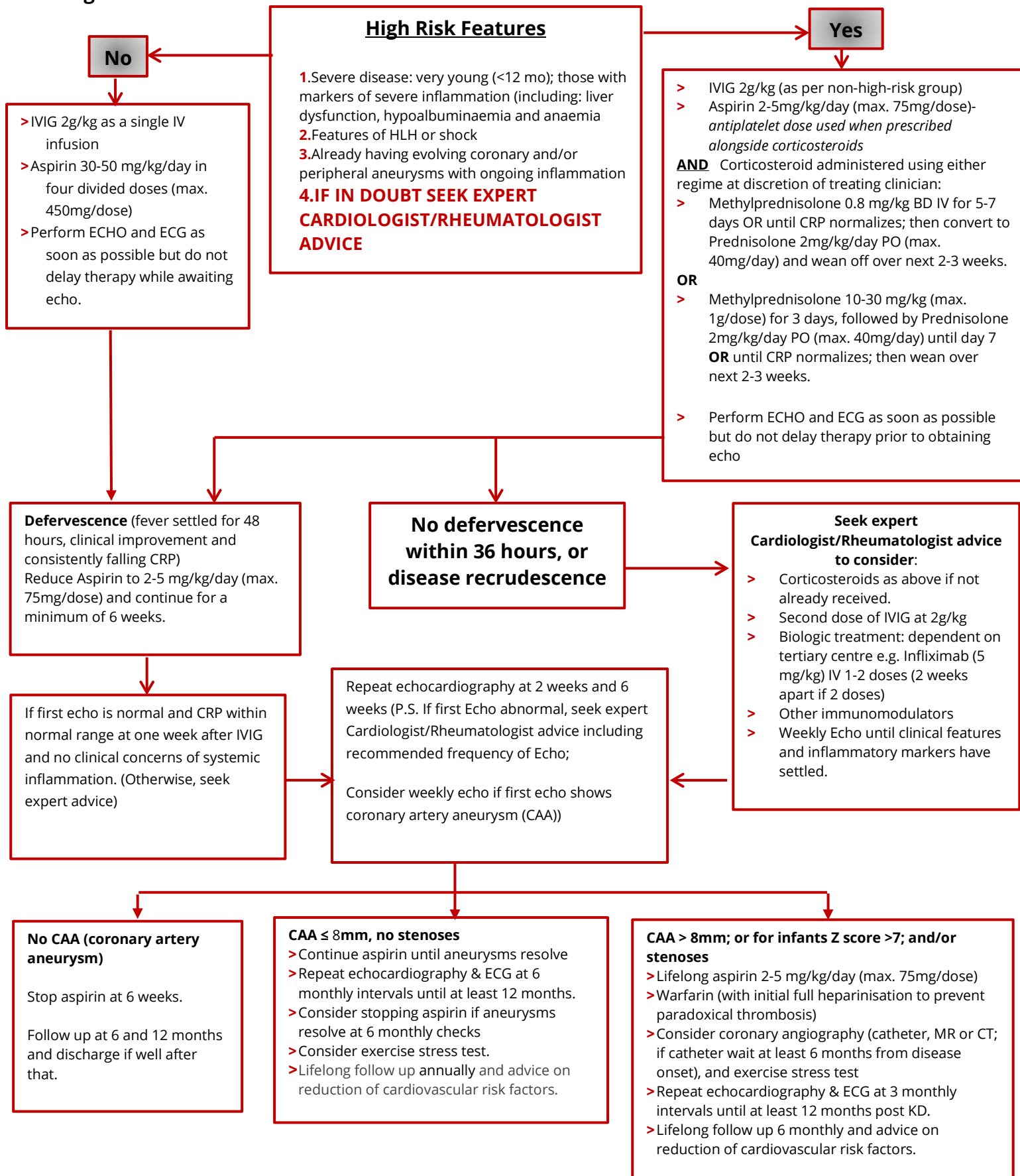
- Review in second week of illness to ensure no symptoms or signs of evolving cardiac failure and to repeat ECG, Echo and review investigation results. Repeat CRP at this week 2 follow up if CRP was not <10mg/L at discharge.
- Repeat echocardiogram in out-patient clinic at 2 weeks and then 6-8 weeks.
- If evidence of coronary artery aneurysm at any stage, refer to cardiologist for intermediate and long term follow up into adult life. Suggested ongoing follow up and imaging:
 - > 6 and 12 month reviews if no coronary artery involvement or dilatation only. If no aneurysms ever identified with normal Echo findings, discharge at 12 months
 - > 6 and 12 month reviews if small or medium aneurysms found and then annual review thereafter
 - > 3 monthly reviews if giant aneurysms found until 12 months and then 6 monthly reviews thereafter.
- Aneurysms identified, plan for long-term follow up/ investigation (see above and flow chart in diagram 2).

Immunisation

- IV immunoglobulin may interfere with the immune response to live vaccine viruses. If protection is not required imminently, delay live vaccinations for at least 3-6 months after IVIG. Thereafter, all vaccines should be administered as per Department of Health schedule. Where rapid protection is required, vaccination should proceed but may require repetition at a later stage to ensure longer term protection.
- There is some evidence that immune response to the MMR vaccine can be attenuated for up to 11 months after receiving IVIG. It should be considered whether to delay the MMR vaccine by a longer period of at least 11 months.
- Children with high risk of exposure to measles can receive their MMR earlier but, re-vaccination/ booster should be planned for a later date.

Treatment of Kawasaki's Disease

Diagram 2



Appendix A- Prescribing summary

Medication	Dosing	Duration	Weaning
<p>IV immunoglobulin (see appendix B for IVIG prescribing advice)</p>	2g/kg as a single dose	<p>Single IV infusion</p> <p>Rates of the infusion are weight and IVIG brand dependent. Refer to local IVIG policy/ contact local pharmacy team who will calculate the recommended rates and duration on patient's ideal body weight</p>	<p>Usually single dose</p> <p>Only repeat if advised on discussion with Rheumatology</p>
<p>Aspirin</p> <p><i>Please note aspirin is not very soluble in water and there is no liquid form available, so it needs to be prescribed as a measurable dose achievable with full, halves or quarters of the available dispersible tablets.</i> <i>Available formulations are 75mg and 300mg dispersible tablets.</i></p>	<p>Acute febrile phase: 7.5-12.5mg/kg/dose (up to max. 450mg) four times a day</p> <p><i>Round aspirin dose to the nearest measurable unit using quarter or halves of the available tablets (e.g. a 75mg tablet can split to give 37.5mg or 18.75mg). Do not try to disperse in water to try to achieve a proportional dose.</i></p> <p>In the rare case of neonatal Kawasaki disease, aspirin doses would differ for neonates- see BNFC</p>	<p>Continue until afebrile for 48hours, with clinical features improving and CRP falling, then reduce to antiplatelet dose</p> <p><i>Please step down to the antiplatelet dose of aspirin if corticosteroids are commenced</i></p>	<p>Reduce to antiplatelet dose of 2-5mg/kg/dose (max 75mg) once daily and continue for minimum of 6 weeks until the 6-8 week follow up ECHO</p> <p>Aspirin can be stopped if the repeat ECHO at 6-8 weeks is normal and aneurysms never seen at any stage. If aneurysms seen/ remain, it must be continued - see final boxes of diagram 2</p>
<p>Corticosteroids regimen 1* (discuss choice of regimen with Rheumatology)</p>	IV Methylprednisolone 0.8mg/kg IV BD	5-7 days or until CRP normalises	Then convert to oral prednisolone 2mg/kg/day (maximum 40mg) and wean slowly over next 2-3 weeks
<p>Corticosteroids regimen 2* (discuss choice of regimen with Rheumatology)</p>	IV Methylprednisolone 10-30mg/kg (maximum of 1gram/day) once daily	3 days	Then convert to oral prednisolone 2mg/kg/day (maximum 40mg) for further 4 days or until CRP normalises, then wean slowly over next 2-3 weeks.
<p>Proton Pump Inhibitor (PPI)</p>	As per BNF	A PPI should be prescribed whilst patient is on acute phase aspirin and especially if they are also on corticosteroids	

Appendix B- IVIG Tips

Brand of IVIG

For patients who have never received IV immunoglobulin before, any brand of immunoglobulin could be used but choice may depend on availability so always discuss with pharmacy. For patients who have previously received IV immunoglobulin they should be prescribed the same brand as previously given. Discuss all prescriptions/ requests for IV immunoglobulin with pharmacy including out of hours.

Approval

IV immunoglobulin panel approval is **not** needed for use in Kawasaki Disease as it is already an approved indication.

Prescribing and vial choice

If prescribing using an electronic prescribing system that requires vial strength to be selected, choose the most cost-effective way to prescribe bearing in mind vial strengths to avoid unnecessary wastage. For example, for a dose of 30 grams, one should prescribe a 10gram vial and a 20gram vial separately.

Obesity and large doses

Dosing should be based on Ideal Body Weight (IBW) for obese patients. For any patient, if the calculated dose of 2g/kg comes to more than 80 grams, consider splitting the dose over 2 days to avoid fluid overloading the patient. **When splitting the dose, consider doing so to minimize wastage from vials, for example:** for a total dose of 90g, rather than prescribing 45grams on day 1 and 45grams on day 2, you could prescribe 50grams on day 1 and 40grams on day 2.

Fluid Restricted Patients

Advice is to split the IV immunoglobulin dose of 2g/kg over 2 or more days. Alternatively, careful use of a one-off dose of furosemide could be considered, but discuss this first with the regional paediatric Rheumatology and Renal teams.

Administration

Nursing staff should document the batch number in the patient notes. Rates of administration increase gradually and will be provided by pharmacy. Rates are formulation specific.

Intention

The aim of IVIG treatment is to switch off the inflammatory process and timely treatment with IVIG reduces the risk of coronary artery aneurysms by >75%. Delays to effective immunomodulatory treatment, including IVIG, increase the risk of coronary artery aneurysms.

Repeat IVIG dose?

In cases of refractory Kawasaki disease i.e. those in which fever fails to resolve and CRP does not rapidly fall within 36-48hours of the first IVIG dose, a 2nd dose of IV immunoglobulin might be considered. This decision **must** first be discussed with Rheumatology. In a patient who has shown some but not complete response to the first dose, second IVIG dose may be appropriate at the same

time as starting corticosteroids. A second dose of IVIG is unlikely to be beneficial if there was little response to the first dose.

Research

Clinicians managing KD are encouraged to access clinical trials where available. Clinical teams in tertiary centres may be able to signpost if appropriate.

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