

### Kawasaki Disease (PIP)

<b>Key Document code:</b>	WAHT-TP-062	
<b>Key Documents Owner:</b>	Dr V Weckemann	Consultant Paediatrician
<b>Approved by:</b>	Paediatric Quality Improvement meeting	
<b>Date of Approval:</b>	9 <sup>th</sup> March 2024	
<b>Date of review:</b> This is the most current document and should be used until a revised version is in place	9 <sup>th</sup> March 2027	

<b>Date</b>	<b>Amendment</b>	<b>Approved by:</b>
9 <sup>th</sup> March 2024	Document reviewed and amendments included from updated PiP guideline 2022-2024	Paediatric Governance Meeting

**The following guidance is taken from the Partners In Paediatrics (PIP)**

Kawasaki 2022–24

# KAWASAKI DISEASE

*Early treatment reduces mortality from coronary artery aneurysms*

## RECOGNITION AND ASSESSMENT

### Symptoms and signs

- Fever  $\geq 5$  days and 4 of the following:

Feature	Details
conjunctivitis	bilateral, bulbar, non-exudative
oral changes	red lips/pharynx/tongue
peripheral oedema	erythema palms and soles, followed by desquamation fingertips 10–15 days after onset of fever
rash	polymorphous (no vesicles or crusts)
lymph nodes	acutely enlarged cervical nodes $>1.5$ cm diameter

- Evidence of an infectious trigger does not exclude Kawasaki disease
- Presence of a coronary artery aneurysm with any 1 of the above features is diagnostic

### Other features

- Most common in children aged  $<5$  yr, peak 18–24 months
- Atypical cases may not fulfil all the above criteria
- if fever  $<5$  days but 4 signs above
- persistent raised CRP and no other diagnosis and suspicion of Kawasaki disease (KD)
- fever usually precedes the other signs, unresponsive to antipyretics
- common features: irritability, erythema of BCG site
- other features include aseptic meningitis, uveitis, cough, vomiting, diarrhoea, abdominal pain, urethritis, arthralgia and arthritis
  - examine for aneurysms in other areas e.g. axillary

### High risk features

- Already failed IVIG
- Aged  $<1$  yr
- Severe inflammation (persistently raised CRP despite IVIG, liver dysfunction, hypoalbuminaemia, anaemia)
- Features of haemophagocytic lymphohistiocytosis (persistent fever, hepatosplenomegaly, cytopenia  $>2$  cell lines, hypertriglyceridaemia, hypofibrinogenaemia, increased D-dimers, hyperferritinaemia, failing ESR)
- Shock
- Evolving coronary or peripheral aneurysms
- Kobayashi risk score  $>5$

Parameter	Score
Na $\leq 133$ mmol/L	2
$\leq 4$ days of illness	2
ALT $\geq 100$ iu/L	1
platelets $\leq 300 \times 10^9/L$	1
CRP $\geq 100$ mg/L	1
aged $\leq 1$ yr	1
$\geq 80\%$ neutrophils	2

### Investigations

None is diagnostic

- FBC: neutrophilia and thrombocytopenia early
- ESR and CRP elevated
- LFTs: raised bilirubin, ALT, low albumin
- Urine: sterile pyuria (proteinuria is suggestive of an alternative diagnosis)
- CSF: lymphocytes

Please note that clinical key documents are not designed to be printed, but to be viewed on-line. This is to ensure that the correct and most up to date version is being used. If, in exceptional circumstances, you need to print a copy, please note that the information will only be valid for 24 hours

- ECG: ST depression, T wave inversion, heart block
- Echo: do not delay therapy before echocardiogram
- Throat swab for Group A strep
- Anti-streptolysin O titre (ASOT) or anti-DNase B for evidence of streptococcal infection
- Blood culture
- Urinalysis, microscopy and culture
- If rash present, serology for enterovirus, parvovirus, EBV, CMV; if features of measles, urine or throat swab in viral transport medium for PCR

### Incomplete Kawasaki disease

- Children with fever  $\geq 5$  days and 2 or 3 compatible clinical criteria **or**
- Infants with fever  $\geq 7$  days with other explanation
- CRP  $< 30$  mg/L and ESR  $< 40$  mm/hr
  - if fever persists, serial clinical and laboratory re-evaluation
  - if typical peeling develops, echocardiogram
- CRP  $\geq 30$  mg/L and/or ESR  $\geq 40$  mm/hr treat if:
  - anaemia for age
  - platelets  $\geq 450 \times 10^9/L$  after 7<sup>th</sup> day of fever
  - albumin  $< 30$  g/L
  - elevated ALT
  - WBC  $> 15 \times 10^9/L$
  - urine  $\geq 10$  WBC/microlitre

## IMMEDIATE TREATMENT

- Aspirin 7.5–12.5 mg/kg oral 6-hrly until afebrile or a minimum of 2 weeks
- Intravenous immunoglobulin (IVIG) 2 g/kg
  - check concentration (g/mL) for preparation used in your Trust
  - administer at gradually increasing rate, as below:

Rate*	Duration
30 mg/kg/hr	30 min
60 mg/kg/hr	30 min
120 mg/kg/hr	30 min
240 mg/kg/hr*	30 min
360 mg/kg/hr*	30 min
480 mg/kg/hr*	To completion

\* Volume will depend on concentration used and maximum rate may be restricted by product literature

**Start IVIG as soon as possible (delayed treatment increases risk of aneurysm)**

## MONITORING IVIG INFUSION

- Monitor temperature, heart rate, BP and respiratory rate:
  - every 5 min for first 15 min
  - then every 15 min for first hour
- Anticipate anaphylaxis, flushing, fever, headache, shivering
- If tolerated, increase infusion rate to give total dose over remaining 10 hr and monitor hourly
- If mild reaction, stop infusion for 15 min then restart at slower rate

## HIGH RISK

- Aspirin and IVIG as above
- Methylprednisolone 0.8 mg/kg IV 12-hrly for 5–7 days or until CRP normalises
  - then prednisolone 2 mg/kg/day oral and wean over 2–3 weeks

## SUBSEQUENT MANAGEMENT

- If fever persists 36 hr after completion of IVIG, consider a single repeat dose of IVIG (as above)
- If fever persists after second dose IVIG give methylprednisolone IV as above if not already given
- Discuss with cardiologist about infliximab (6 mg/kg) IV 1–2 doses (2 weeks apart if 2 doses)
- Fever settled for 48 hr, clinical improvement and falling CRP, reduce dose of aspirin to 2–5 mg/kg (maximum 75 mg) oral as single daily dose for minimum 6 weeks (until result of echocardiogram known)

## DISCHARGE AND FOLLOW-UP

- Discharge when fever settles
- Echocardiogram at 10–14 days and 6 weeks from onset of signs and symptoms
- Outpatient appointment 1 week after echocardiogram
- Advise to avoid excessive strenuous activity until outpatient appointment after echocardiogram
- Advise to avoid all live vaccines (e.g. MMR) for 3 months following IVIG therapy

## OUTPATIENT MANAGEMENT

- No aneurysms at 6 weeks echocardiogram
- stop aspirin
- no restriction on activity
- follow-up at 12 months and discharge if well
- Single aneurysm <8 mm diameter
- aspirin 2–5 mg/kg (maximum 75 mg) once daily until aneurysm disappears
- cardiologist will advise on limitation of activity, exercise stress test, MR/CT angiogram
- 6-monthly ECG and echocardiogram
- lifelong follow-up and advice on reduction of cardiovascular risk factors
- Multiple or giant aneurysm or stenosis
- as for single aneurysm **and**
  - lifelong aspirin 2–5 mg/kg/day
  - warfarin (after heparinisation)