Diagnosis and Management of Kawasaki Disease

Reference: 1507
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1. Introduction
Kawasaki disease should be considered in any child with prolonged fever lasting > 5 days. It most commonly occurs in young preschool children but can occur in all age groups including adults and infants. It may be more difficult to diagnose outside the normal age range. The cause is unknown but without appropriate treatment 15-25% of children will develop coronary artery aneurysms. Without treatment fever may settle by 10-11 days but may persist for several weeks.

2. Guideline Content

A. DIAGNOSIS (I)
Classical diagnosis is based on fever for > 5 days and 4 of 5 principal features:

**Changes in extremities** - early: erythema palms/soles, oedema hands/feet
Week 2-3: periungual peeling fingers and toes

**Rash**

**Non exudative conjunctivitis**

**Lip and mouth changes**: redness, cracking, strawberry tongue

**Cervical adenopathy** (usually unilateral > 1.5cm)

All these features may not be present at the one time so diagnosis also relies on a history of any of these features during the illness.

**Laboratory findings:**
Blood results are not part of the classical diagnostic criteria but can be helpful
ESR / CRP: usually elevated
Platelet count: elevated (>450 by day7*)
Raised WBC count (>15.00)
Abnormal LFT
Hypoalbuminaemia
Sterile pyuria
Aseptic meningitis

**Imaging:**
A baseline Echo is recommended as aneurysma and mycocarditis may occur early in the disease course. An ECHO should also be routine in most children with PUO

**Diagnosis (ii) Incomplete or atypical disease**
Since Kawasaki disease was first described some children have been found to have coronary artery abnormalities without having had an illness that fulfils all the criteria. This is more common in young infants or adults who may not even have the classical unilateral cervical adenopathy. Revised clinical criteria include children with 2 or 3 clinical criteria, fever >5 days, raised CRP/ESR and 3 of the above laboratory criteria as being possible Kawasaki disease. Early ECHO is also recommended in this group which may help with the diagnosis although it is unusual to see coronary artery changes before 10 days. Occasionally infants and children will present with thrombocytopenia which is associated with a poor prognosis and delayed identification.

See Guidance for Paediatric multisystem inflammatory syndrome temporally associated with Covid-19

B. DIFFERENTIAL AND MISSED DIAGNOSES

A number of other conditions may mimic Kawasaki disease:

- **Viral infections:** EBV, Measles, adenovirus, enteroviruses
- Serum samples (EBV), saliva (measles) and stool and throat swabs (adeno/entero) may be useful
– Scarlet fever: some children may however have both Kawasaki disease and evidence of group A strep
– Staphylococcal scalded skin
– Toxic shock syndrome
– Systemic Juvenile rheumatoid arthritis
– Drug reactions
– Stevens-johnsons
– Leptospirosis
– Missed diagnoses:

A child with unilateral cervical adenopathy and fever who is given antibiotics who develops a skin rash may be labelled as a drug reaction
Fever, rash and culture negative CSF pleocytosis may be felt to be viral meningitis
Occasionally child may present with fever & an apparent acute abdomen or hepatitis

C. TREATMENT

1. **Immunoglobulin**: IVIG 2g/kg given within 10 days of fever developing, significantly reduces the risk of coronary artery abnormalities
   This can be given usually over 12 hours but in the presence of cardiovascular compromise it may be appropriate for the dose to be split to give 1g/kg on 2 consecutive days. The trust guidelines on immunoglobulin infusions should be followed.
   As Kawasaki disease is a RED indication for immunoglobulin it should easily obtained form pharmacy if the appropriate form is completed by the consultant in charge, or under their direction. To obtain Immunoglobulin out of hours please speak to the on call pharmacist.
2. Please always check Asprin doses against BNFC
3. **Asprin**: High dose asprin 30-50 mg/kg/ day in 4 divided doses till fever has settled.Followed by 3-5mg/kg/day till repeat ECHO in 6-8 weeks
4. Where the clinical diagnosis is very likely and treatment has not been given beyond 10 days of fever intravenous immunoglobulin is still indicated if the child is still febrile or still has raised inflammatory makers
5. Any child with documented cardiac lesions, coronary artery aneurisms or signs of myocarditis at diagnosis should be discussed acutely with the Paeditairc cardiologists in Leeds
6. In a child who has relapse of fever where Kawasaki disease is felt clinically likely, a further dose of IVIG should be given but alternative diagnoses should also be considered
7. **Steroids**: while high dose methylprednisolone has not been shown to be of additional benefit as a routine treatment in Kawasaki disease, lower dose prednisolone has been shown to be of benefit in relapsed Kawasaki and may be considered in children with severe Kawasaki disease at presentation including those under 1 year of age or with documented aneurisms at presentation. Methylprednisolone 0.8mg/kg b.d for 5-7 days followed by 2-3 weeks of oral prednisolone 2mg/kg has been recommended in a recent UK review article*
8. **Infliximab, cyclosorin and cyclophosphamide** have been used in refractory patients but this must be done in conjunction with the Rheumatology or Immunology/ ID team

E. FOLLOW UP
All children with possible Kawasaki disease need referral to the cardiologists to discuss the indication for follow up by them or by the local paediatrician at 6-8 weeks. Any child with documented cardiac problems with possible coronary artery involvement or myocarditis should be discussed with cardiology at the time.

There is controversy about the need for long term follow up of children who have had no documented cardiac lesion. Children who have had cardiac involvement will generally be followed by the cardiologists in their Sheffield clinic.

Any child with recurrent (a second episode at a different time,) or relapsed Kawasaki disease (where the initial fever recurs or does not settle) should be referred to the Immunology/ID or rheumatology team.

3. References

American Heart Association Guidelines 2004
Eleftheriou et al Arch Dis Child 2014;99:74-83