Kawasaki Disease

Management Guidelines
Emergency Department
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Kawasaki disease is a common vasculitis of childhood especially in < 5yo.

Self-limiting, with fever & manifestations of acute inflammation lasting 12 days (average) without therapy.

Complications include coronary artery aneurysms, depressed myocardial contractility & heart failure, myocardial infarction, arrhythmias and peripheral arterial occlusion.

- Infants under 12 months at increased risk of coronary artery aneurysm
- Delay of treatment (after 10 days) increases risk of coronary artery aneurysm by 5X

Diagnostic criteria

Presence of prolonged *unexplained* fever ≥ 5 days (fever >38.5C)
with at least 4/5 following criteria

1. Bilateral non-exudative conjunctivitis
2. Polymorphous rash
3. Cervical lymphadenopathy (at least 1 LN >1.5cm in diameter)
4. Mucositis-cracked red lips, injected pharynx or strawberry tongue.
5. Extremity changes-erythema of palms/soles, oedema of hands/feet (acute phase), and periungual desquamation (convalescent phase)

Oedema of hands

Rash

Desquamation of fingers

Associated non-specific symptoms

- Diarrhoea,vomiting ,or abdominal pain (60%)
- ***Irritability*** (50%)
- Joint pain (15%)
- Weakness (19%)

Incomplete KD-do not fulfil diagnostic criteria (<4 signs of mucocutaneous inflammation) but other wise similar clinical picture to that of "classic" KD. Infants(<12/12) and >5yo more likely to have incomplete. Still at risk of cardiovascular sequelae **So if prolonged unexplained fever please consider and discuss with ED consultant/DPAM on-call consultant**.

Cracked lips and conjunctival injection

Alternative Diagnosis

- Measles,adenovirus,EBV
- Scarlet fever, Toxic Shock Syndrome
- Steven-Johnson Syndrome

Lab findings (not diagnostic but supportive)

- Elevated acute phase reactants (CRP,ESR)
- Elevated WCC with predominant neutrophilia
- Elevated platelets (after 1 week)
- Normocytic, normochromic anaemia
- Urethral pyuria (need clean voided specimen)

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Initial Investigations:

CRP ,ESR ,FBP,ALT,Albumin
ASOT/AntiDNAase B
Urinalysis-preferably clean catch-micro examination
Blood culture

All suspected cases should be discussed with the ED Admitting Registrar/Consultant for admission under DPAM on-call Consultant

Ongoing care

Referral to Cardiology by DPAM for echocardiogram only after the diagnosis of KD made/confirmed and treatment instituted. Many patients (esp. those < 3 years) will require sedation to perform the echocardiogram as the irritability (so commonly seen) precludes performing adequate echocardiogram in the acute phase. Echocardiogram is required at/after initial diagnosis and repeated again at 4-8 weeks post treatment. Echocardiography plays no role in the diagnosis of KD.
Referral to Infectious disease consultant is at the discretion of the DPAM Consultant.

Initial Treatment

IV Immunoglobulin (IVIG) 2gm/kg over 6-12 hours
Low dose aspirin at 3-5mg/kg daily
Low threshold for 2nd dose IVIG if incomplete treatment response, particularly in high risk age groups and "atypical" KD

Discharge Treatment

Continue aspirin (low dose) at 3-5 mg/kg daily continued until repeat echo at 4-8 confirms absence of coronary involvement