

KAWASAKI DISEASE

Introduction

A systemic febrile condition affecting children usually < 5 years old.

Aetiology remains unknown, possible bacterial toxins or viral agents with genetic predisposition. Also known as mucocutaneous lymph node syndrome.

Table 1. Diagnostic criteria for Kawasaki Disease

Diagnostic Criteria
<ul style="list-style-type: none">• fever lasting at least 5 days.• at least 4 out of 5 of the following:<ul style="list-style-type: none">- bilateral non-purulent conjunctivitis- mucosal changes of the oropharynx (injected pharynx, red lips, dry fissured lips, strawberry tongue).- changes in extremities (oedema and/or erythema of the hands or feet, desquamation, beginning periungually).- rash (usually truncal), polymorphous but non vesicular.- cervical lymphadenopathy• illness not explained by other disease process.

Clinical Pearls

Diagnosis is via criteria listed in Table 1.

Other helpful signs:

- indurated BCG scar
- perianal excoriation, irritability
- altered mental state
- aseptic meningitis
- transient arthritis
- diarrhoea, vomiting, abdominal pain
- hepatosplenomegaly
- hydrops of gallbladder
- sterile pyuria

Investigations

- full blood count
 - anaemia, leucocytosis, thrombocytosis.
- ESR and CRP elevated
- serum albumin < 3g / dl; Raised alanine aminotransaminase
- urine >10 wbc / hpf
- chest X-ray, ECG.
- echocardiogram in the acute phase and repeat at 6-8 weeks or earlier if indicated.

Most important complication is coronary vasculitis, usually within 2 weeks of illness, affecting up to 25% of untreated children. Usually asymptomatic, it may manifest as myocardial ischaemia, infarction, pericarditis, myocarditis, endocarditis, heart failure and arrhythmias.

Incomplete Kawasaki Disease

Patients who do not fulfill the classic diagnostic criteria outlined above. Tends to occur in infants and the youngest patients. High index of suspicion should be maintained for the diagnosis of incomplete KD. Higher risk of coronary artery dilatation or aneurysm occurring.

Echocardiography is indicated in patients who have prolonged fever with:

- two other criteria,
- subsequent unexplained periungual desquamation,
- two criteria + thrombocytosis
- rash without any other explanation.

Atypical Kawasaki Disease

For patients who have atypical presentation, such as renal impairment, that generally is not seen in Kawasaki Disease.

Treatment

Primary treatment

- IV Immunoglobulins 2 Gm/kg infusion over 10 - 12 hours.
Therapy < 10 days of onset effective in preventing coronary vascular damage.
- Oral Aspirin 30 mg/kg/day for 2 weeks or until patient is afebrile for 2-3 days.

Maintenance: Oral Aspirin 3-5 mg/kg daily (anti-platelet dose) for 6 - 8 weeks or until ESR and platelet count normalise.

If coronary aneurysm present, then continue aspirin until resolves.
Alternative: Oral Dipyridamole 3 - 5 mg/kg daily

Kawasaki Disease not responding to Primary Treatment

Defined as persistent or recrudescence fever \geq 36 hours after initial IVIG infusion.

Treatment: Repeat IV Immunoglobulins 2 Gm/kg infusion over 10 - 12 hours

Table 2. Risk stratification and long term follow up

risk level	treatment	physical activity	follow up	invasive testing
Level I no coronary artery changes	none beyond 6-8 weeks	no restrictions beyond 6-8 weeks	cardiovascular risk assessment, counselling at 5yr intervals	none
Level II transient coronary artery ectasia; none after 6-8 weeks	none beyond 6-8 weeks	no restrictions beyond 6-8 weeks	cardiovascular risk assessment, counselling at 3 to 5yr intervals	none
Level III one small-medium coronary artery aneurysm, major coronary artery	low dose aspirin until aneurysm regression documented	age <11 yr old: no restriction beyond 6-8 weeks . avoid contact sports if on aspirin	annual echocardiogram and ECG, and cardiovascular risk assessment counselling	angiography if non-invasive test suggests ischemia
Level IV > 1 large or giant coronary artery aneurysm, or multiple or complex aneurysms in same coronary artery, without destruction	long term aspirin & warfarin (target INR 2.0-2.5) or LMWH in giant aneurysms	avoid contact sports	biannual echocardiogram and ECG; annual stress test	angiography at 6-12 mo or sooner if indicated; repeated study if non-invasive test, clinical or laboratory findings suggest ischemia
Level V coronary artery obstruction	long term aspirin; warfarin or LMWH if giant aneurysm persists. also consider β -blockers	avoid contact sports	biannual echocardiogram and ECG; annual stress test	angiography to address therapeutic options

LMWH, low molecular weight heparin

Vaccinations

The use of Immunoglobulins may impair efficacy of live-attenuated virus vaccines. Delay these vaccinations for at least 3-6 months.

Prognosis

Complete recovery in children without coronary artery involvement.

Most (80%) 3 - 5 mm aneurysms resolve; 30% of 5 - 8 mm aneurysms resolve.

Prognosis worst for aneurysms > 8 mm in diameter. Mortality in 1 - 2 %, usually from cardiac complications within 1 - 2 months of onset.