

Idiopathic systemic disorder predominantly of young children described by Tomisaku Kawasaki in 1967 that is characterised by fever, cutaneous/mucosal changes and vasculitis of small & medium blood vessels. Most common cause of acquired heart disease in developed countries.

### Epidemiology

- Highest incidence is in Asians esp Japan. ~10-20x less common in Caucasians.
- 85% affected children are under 5 years old. Uncommon over the age of 8.
- 1.5M:1F.
- Occasional local epidemics more common in Winter and Spring.

### Aetiology

Currently unknown. Epidemiology supports a possible infective agent - e.g. a bacteria (Strep or Staph) producing a toxin that acts as a superantigen binding T-cell receptor to the MHC II of an antigen presenting cell initiating & prolonging the inflammatory response.

### Pathology

- Systemic vasculitis similar to infantile periarteritis nodosa.
- Coronary aneurysms are usually present at autopsy.
- The vasculitis is characterized by acute inflammation with no or mild fibrinoid necrosis.

### Diagnostic criteria

- Fever (generally  $\geq 39.5^{\circ}\text{C}$ ) of unknown origin for  $\geq 5\text{d}$
- And 4 from 5 of:
  - Lip & oral changes (lip fissures, pharyngitis, strawberry tongue) [90%]
  - Bilateral dry bulbar conjunctivitis (with limbic sparing) [85%]
  - Polymorphous rash (trunk, perineum, limbs) [80%]
  - Extremity changes (redness/swelling/late desquamation) [75%]
  - Cervical lymph nodes  $>1.5\text{cm}$  (usually unilateral) [40%]
- Atypical/incomplete (not fulfilling enough criteria) in ~30% - especially in infants

### Clinical features

Phase	Time from fever onset	Features
Acute	1-2 weeks	Highly febrile Irritable and toxic-appearing Oral changes rapidly follow Oedema and erythema of feet Rash especially common in perineal area
Subacute	2-8 weeks	Gradual improvement Fever settles Desquamation of perineum, palms, soles Arthritis, arthralgia Thrombocytosis Coronary artery aneurysm Myocardial infarction
Convalescent	Months to years	Resolution of remaining symptoms Laboratory values return to normal Aneurysms may resolve or persist Beau's lines Cardiac dysfunction and MI may still occur

### Subsidiary features:

- Cardiovascular: pancarditis, aortic or mitral incompetence
- Respiratory: pneumonitis, coryzal, otitis media
- Gastrointestinal: hydrops of gallbladder, jaundice, diarrhoea
- Blood: mild anaemia, thrombocytosis
- GUS: sterile pyuria, mild proteinuria, nephritis, priapism
- CNS: aseptic meningitis, cranial nerve palsies
- Musculoskeletal: arthritis, arthralgia
- Other: anterior uveitis, BCG scar inflammation

### Investigations

Urinalysis: sterile pyuria and mild proteinuria.

Bloods: WBC, platelets, liver transaminases, bilirubin, ESR and CRP may be ↑. Hb & albumin ↓

ECG: Various anomalies (prolonged PR or QT, ↓R amplitude, ST↓, T wave flat/inverted)

Imaging: CXR (Signs of heart failure), Echo (LV fn, valves, coronary), USS abdo (GB), stress testing (ischaemia), angiography or MRI/MRA: aneurysm and thrombus size

### Differential diagnosis

- Bacterial infections (streptococcal scalded-skin syndrome, TSS, scarlet fever)
- Viral illnesses (Measles, Roseola, Rubella, EBV, Enterovirus etc)
- Toxoplasmosis
- Rocky Mountain spotted fever
- Leptospirosis
- Acrodynia (mercury toxicity)
- Gianotti-Crosti syndrome
- Collagen vascular diseases, juvenile rheumatoid arthritis
- Drug reactions (e.g. erythema multiforme)

### Treatment

- IVIG 2g/kg over 10hrs within 10d of onset. Dose can be repeated if necessary (20-25%).
- Aspirin: 10mg/kg tds po until defervesce (optional) then 3-5mg/kg/day od po for 2+ months as antithrombotic
- Corticosteroids may speed up defervescence but ?don't improve Cx. Used if IVIG fails.
- Bed rest for 2 to 3 weeks
- Follow up echocardiograms
- Delay of live immunisations for 9mo after IVIG
- PTCA / Thrombolysis / CABG / Cardiac transplantation have been used on occasion

### Complications and Prognosis

- ~20-25% of untreated → coronary aneurysms. Reduced to <5% with prompt IVIG Rx.
- Aneurysms more likely in males, age <1 yr or >8 yrs, if fever >14/7, or init blds abnormal.
- Most (~90%) aneurysms regress by 2 years
- Bowel/peripheral ischaemia leading to gangrene
- Reye's Syndrome from aspirin use (advised to stop aspirin if get varicella or influenza)
- Recurrence is rare (<1-3%)
- Mortality:
  - 4% in infants to <<1% in older children.
  - Mainly due to thrombosis of coronary artery aneurysms → MI or myocarditis
  - Mortality greater in giant aneurysms (>8mm)