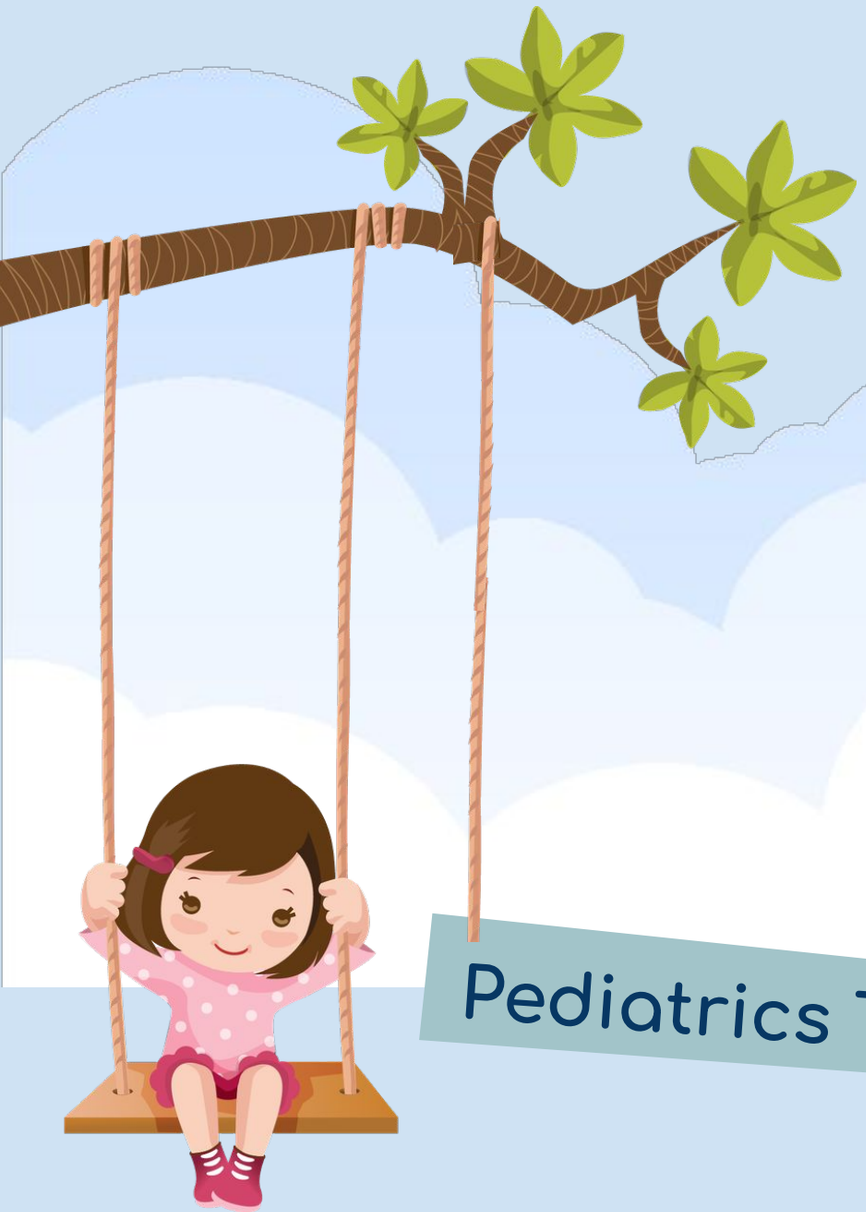

Pediatrics TeamWork

437

Kawasaki disease

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Kawasaki disease topic includes also dr. Mohammed alghamdi slides and notes from acquired heart disease lecture to avoid repeating topics!

Notes Previous Notes Book Important!

Kawasaki Disease

Overview:

- The other name is mucocutaneous lymph node syndrome
- Kawasaki disease mainly affects children of 6 months to 4 or 5 years of age, with a peak at the end of the first year of life.
- **It is the commonest cause of acquired heart disease in children** (in north america) (in SA is Acute Rheumatic Fever):

Cardiac involvement
Dilation and aneurysm formation, thrombus formation, fibrosis and stenosis, myocardial infarction and it may cause myocarditis and endocarditis

- Systemic inflammatory process (Vasculitis: medium size , mainly coronary arteries) with no known etiology
- Maybe infectious etiology
- More common in children of Japanese and, to a lesser extent, Black-Caribbean ethnicity, than in Caucasians

Diagnosis:

There is no diagnostic test; instead, the diagnosis is made based on clinical findings alone:

Fever > 5 days

At least **4** of the following:

1 Changes in the extremities:
- Erythema and edema of hands and feet (acute phase)
- Subsequent peeling of distal ends of digits (subacute phase)

2 Polymorphous rash. Any type of skin rash except vesicles and bullae. Check the diaper area

3 Non-purulent bilateral conjunctivitis

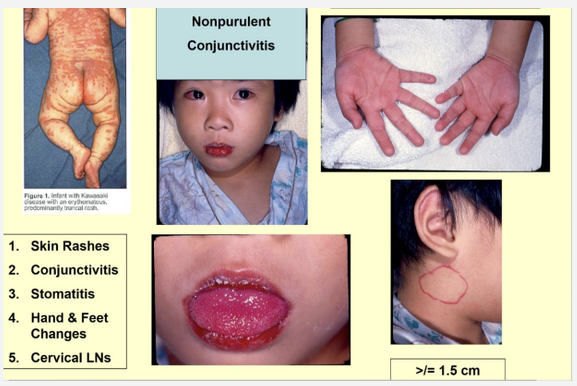
4 Mucosal changes:
- Strawberry tongue
- Red, cracked lips and/or erythema of oral and pharyngeal mucosa

5 Cervical lymph node (>= 1.5 cm in diameter)

-It is NOT a must to have all the clinical features in the same time
-If you don't ask about red eyes the parents may forget about it (it disappears quickly)
-Other Ddx of strawberry tongue : scarlet fever
-Peeling of the skin usually not in the beginning of disease
-**Other medium size arteries:** axillary, femoral, iliac and renal arteries

Kawasaki Disease

Diagnosis & DDX:



Differential diagnoses

- Scarlet fever
- EBV infection
- Adenovirus infection
- Staphylococcal scalded skin syndrome
- Drug reactions
- Stevens-Johnson syndrome

Young infants may have 'incomplete' symptoms or diseases, in which not all the cardinal features are present:

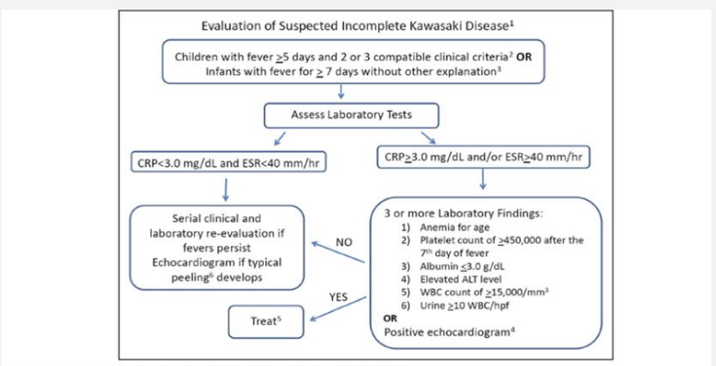


Figure 3. Evaluation of suspected incomplete Kawasaki disease.
(1) In the absence of a "gold standard" for diagnosis, this algorithm cannot be evidence based but rather represents the informed opinion of the expert committee. Consultation with an expert should be sought any time assistance is needed. (2) Clinical findings of Kawasaki disease are listed in Table 3. Characteristics suggesting that another diagnosis should be considered include exudative conjunctivitis, exudative pharyngitis, ulcerative intraoral lesions, bullous or vesicular rash, generalized adenopathy, or splenomegaly. (3) Infants ≤6 months of age are the most likely to develop prolonged fever without other clinical criteria for Kawasaki disease; these infants are at particularly high risk of developing coronary artery abnormalities. (4) Echocardiography is considered positive for purposes of this algorithm if any of 3 conditions are met: *Z* score of left anterior descending coronary artery or right coronary artery ≥2.5; coronary artery aneurysm is observed; or ≥3 other suggestive features exist, including decreased left ventricular function, mitral regurgitation, pericardial effusion, or *Z* scores in left anterior descending coronary artery or right coronary artery of 2 to 2.5. (5) If the echocardiogram is positive, treatment should be given within 10 days of fever onset or after the tenth day of fever in the presence of clinical and laboratory signs (C-reactive protein [CRP], erythrocyte sedimentation rate [ESR]) of ongoing inflammation. (6) Typical peeling begins under the nail beds of fingers and toes. ALT indicates alanine transaminase; and WBC, white blood cells.

For incomplete symptoms, there should remain a high clinical suspicion, **particularly for children less than 6 months of age with prolonged fever** and these children are more likely to develop coronary artery aneurysms which affected children within the first 6 weeks. **It should be treated as complete KD**

Investigations:

- Affected children have **high inflammatory markers** (C-reactive protein, erythrocyte sedimentation rate, white cell count), with a **platelet count that rises typically in the second week of the illness**.
- CBC: Neutropenia, leukocytosis (50%) and nonspecific anemia
- Elevated liver transaminases (40%), low serum albumin level
- Sterile pyuria (33%) , aseptic meningitis (up to 50%)
- **Echocardiography should be performed when the diagnosis is first suspected**, and at 4–6 weeks to identify coronary artery aneurysms; and it may show a pericardial effusion, myocardial disease (poor contractility), endocardial disease (valve regurgitation), or coronary disease with aneurysm formation, which can be giant (>/= 10 or >8 mm in diameter).
    - If the coronary arteries are abnormal, angiography or **magnetic resonance imaging (MRI) will be required**.

Kawasaki Disease

Management:

Risk scores for coronary aneurysm

1. WBC > 12 000
2. Platelet < 350 000
3. CRP > 3+
4. Hct < 3.5
5. Albumin < 3.5
6. Age </= 12 months
7. Male sex

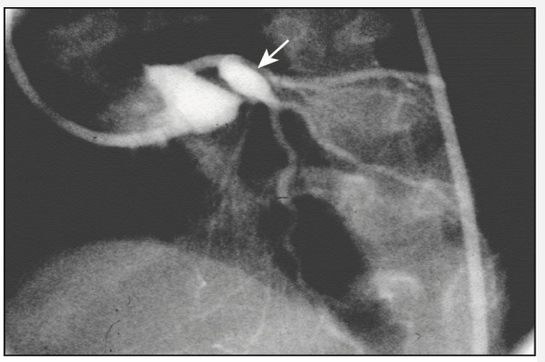


Figure 18.22 Kawasaki disease. Angiogram showing coronary artery aneurysm.

Treatment:

- **Intravenous immunoglobulin** (IVIG), **ideally given within the first 10 days**, to lower the risk of coronary artery aneurysms. From 25% to less than 5%
- **Aspirin** to reduce the risk of thrombosis. due to dilation of the coronary even if there is no dilation start aspirin then re-evaluate after 6 wks with another echo If normal → stop aspirin "coronary changes might develop after this period' , decrease aspirin once afebrile
- **Children with coronary artery aneurysms require long- term low-dose aspirin and lifelong follow-up.**
- Give another anticoagulant if giant aneurysm of CA
- **For resistant Kawasaki disease which presents with fever persists or recurs despite initial treatment: give a second dose of:**
  - intravenous immunoglobulin or,
  - corticosteroids or,
  - infliximab (a monoclonal antibody against tumour necrosis factor-α)

If you find 3/ 6 then treat the pt as Kawasaki disease:
Anemia for age
Plt >450
High WBC count in peripheral blood or in the urine
Low albumin
Elevated Na

Even w/o Tx kids w/ Kawasaki most sx will disappear (like: fever ..). But the problem is w/ coronary involvement:

- Stagnation of flow through the coronaries.
- Dilation of the coronaries.
- Coronary aneurysm.

Whenever you suspect KD or you are not sure, ALWAYS TREAT AS KD!!