



## Kawasaki disease: High index of suspicion needed in a febrile child

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A six-month-old Caucasian boy presents with a three-day history of fever. The parents report that he had red eyes without pus two days previously. On examination, otitis media is noted as well as a nonexudative pharyngitis. Amoxicillin is prescribed. They return on the sixth day of illness and report that the child is still febrile, not responding to antipyretics and has a rash. The rash appears urticarial and the otitis media persists. A different antibiotic is prescribed. The family presents to the emergency room on the eighth day of fever. The child is irritable and difficult to console. He is noted to have red, cracked lips and a nonexudative pharyngitis. He has bilateral otitis media, no lymphadenopathy or conjunctivitis, and has a maculopapular rash on his trunk and extremities. Swelling of the hands and feet or palmar/plantar erythema is not noted. Other than tachycardia, the cardiovascular examination is unremarkable, as is the rest of the examination. Investigations reveal a white blood cell count of  $22 \times 10^9/L$  (normal range  $6 \times 10^9/L$  to  $17.5 \times 10^9/L$ ) with a left shift, hemoglobin level of 90 g/L (normal range 95 g/L to 135 g/L) and platelet count of  $425 \times 10^9/L$  (normal range  $140 \times 10^9/L$  to  $450 \times 10^9/L$ ). His erythrocyte sedimentation rate is 65 mm/h (normal range 3 mm/h to 13 mm/h) and C-reactive protein level is 188 mg/L (normal range 0 mg/L to 5 mg/L). His liver enzyme levels are mildly elevated and albumin level is low. No pyuria is noted. The diagnosis of incomplete Kawasaki disease (KD) is suspected and the child is admitted. He is given intravenous immunoglobulin (IVIG) and acetylsalicylic acid (ASA). Echocardiography reveals a small coronary artery aneurysm.

### LEARNING POINTS

- KD is a systemic vasculitis and the leading cause of acquired heart disease in Canadian children. It can affect children of all ages, but mainly occurs in those <5 years of age.
- This disease is more common among Japanese children and children from the Orient, although it can affect children of all ethnicities.
- The major complication of KD is the development of coronary artery lesions such as dilatations or aneurysms. If untreated, up to 25% of patients will develop coronary artery aneurysms, and mortality may be as high as 2%.
- The diagnosis of KD is not always easy to make because there is no diagnostic test. KD is diagnosed clinically, based on the presence of characteristic clinical findings (Table 1). To establish the diagnosis of classic/complete KD, four of five diagnostic criteria must be present, in addition to fever for  $\geq 5$  days. Furthermore, even patients who do not fulfill criteria (incomplete KD) may develop coronary artery lesions. Therefore, physicians need to have a high index of suspicion for this disease.

**TABLE 1**  
**Diagnostic criteria**

**Classic/Complete Kawasaki disease (KD): Fever for  $\geq 5^*$  days AND at least four of the following clinical criteria:**

- Changes in the peripheral extremities (erythema of the palms and/or soles; edema of the hands and/or feet; or periungual desquamation)
- Polymorphous rash
- Bilateral bulbar conjunctival injection without exudate
- Changes in the lips and oral cavity (erythema and/or cracking of the lips; strawberry tongue; diffuse erythema of the oropharynx)
- Cervical lymphadenopathy (>1.5 cm diameter, usually unilateral)

AND disease not explained by another disease process.

**Incomplete KD: Fever for  $\geq 5$  days with <4 clinical criteria; and disease not explained by another disease process.**

*\*In the presence of >4 clinical criteria, the diagnosis can be made before the fifth day of fever by a clinician experienced in treating KD*

- Very young children, particularly those <6 months of age, and children >9 years of age are more likely to present with incomplete KD. Male sex, extremes of age and prolonged fever are factors associated with a higher risk for the development of coronary artery lesions.
- Other clinical features may be present including irritability, aseptic meningitis, uveitis, gastrointestinal complaints (diarrhea, vomiting, hepatic dysfunction), urethritis/meatitis, hydrops of the gallbladder and arthritis.
- Concomitant infections are common in children with KD. If the infection cannot explain all the clinical features, the diagnosis of KD needs to be entertained.
- Laboratory investigations are nonspecific. Acute phase reactants are usually increased. Other laboratory investigations may be normal, but supportive findings include an elevated white blood cell count with left shift, anemia, thrombocytosis, low albumin level, elevated transaminase levels and sterile pyuria.
- KD is treated with IVIG at a dose of 2 g/kg and ASA at a dose of 80 mg/kg to 100 mg/kg per day in four divided doses. ASA is reduced to an antiplatelet regimen at 3 mg/kg to 5 mg/kg per day after fever has subsided. It is continued until inflammatory markers, platelet count and follow-up echocardiogram at six to eight weeks are normal.
- IVIG is a safe and effective treatment for KD; however, cases of hemolytic anemia have been noted. It is important to be aware of this potential complication and to monitor hemoglobin levels following IVIG administration.

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**RECOMMENDED READING**

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