‘K’ for Kawasaki disease (KD)

What is KD?

Kawasaki disease is an acute medium vessel vasculitis that primarily affects young children. It presents as an acute febrile illness and can lead to development of cardiac complications, if not diagnosed and treated in time.

How common is KD?

Kawasaki disease has emerged as the most common pediatric vasculitis. KD occurs worldwide and affects children of all races, although Asians are believed to be at highest risk. Last reported data from Japan suggests that almost 1 in 100 children in Japan will have KD by the age of 5 years. There is lack of unified epidemiological data on KD from major part of India but several hospitals are now experienced with diagnosis and management of this condition.

What is the etiology of KD?

Even after more than 50 years of its first description, the etiology of KD remains an enigma. Ongoing research has presented evidence indicating that an infectious cause may induce inflammation in individuals genetically predisposed to the condition. Yet, the specific genetic elements responsible for this susceptibility in KD remain unidentified.

What is the pathogenesis of KD?

The abnormal immune response in KD results in widespread inflammation, affecting blood vessels in various organs. The inflammation can damage the walls of blood vessels, leading to the formation of aneurysms and other vascular complications. Coronary artery involvement is a significant concern and can lead to serious cardiovascular problems. The vascular damage in KD can result in thrombosis later in life, precipitating heart complications and, in severe cases, sudden death.

How to make a diagnosis of KD?

Kawasaki Disease is diagnosed purely through clinical evaluation, relying on specific sets of clinical criteria (figure1). The clinical features evolve over a period of time (1-3 weeks) and the entire clinical spectrum may not be seen at any one particular point of time. There is no

laboratory test or marker which is pathognomonic of the condition. Some supportive laboratory findings are listed in table 1.

Figure 1: Diagnostic Criteria For Kawasaki Disease

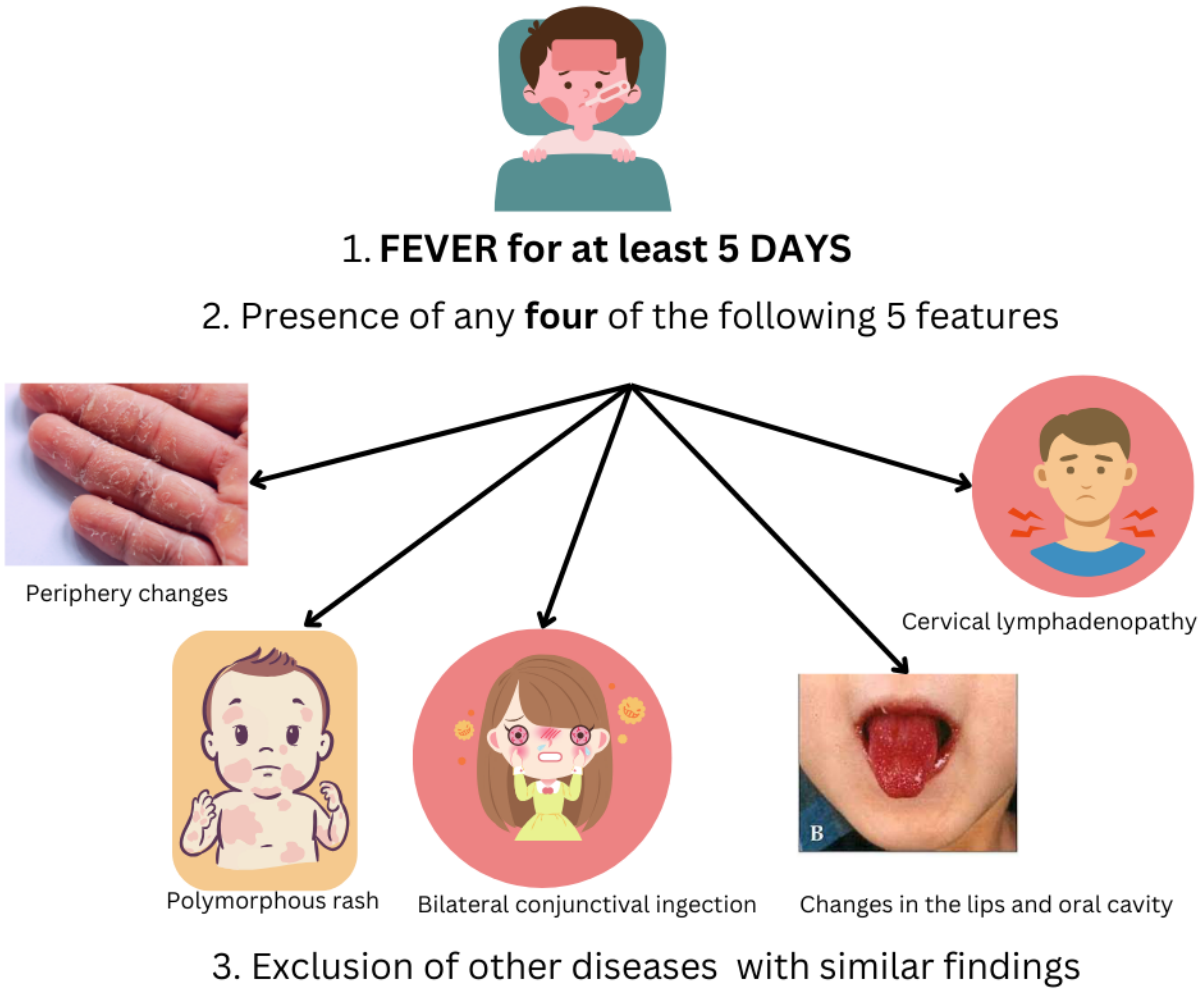

Table 1: Lab findings that may be seen in patients with Kawasaki disease

	Investigation	Findings
1.	Complete blood count	Low hemoglobin, polymorphic leukocytosis and thrombocytosis (in second week)
2.	Inflammatory parameters	High Erythrocyte sedimentation rate, High C-Reactive protein
3.	Markers of cardiac dysfunction	High NT-pro-BNP
4.	Urine routine	Sterile pyuria seen in 30-60% patients
5.	Liver function tests	Transaminitis, hypoalbuminemia

6.	USG abdomen	Gall bladder hydrops
7.	2D echocardiography	Coronary aneurysm, myocarditis, valvulitis, pericardial effusion

What is Atypical/Incomplete KD?

If a patient displays unusual symptoms not typically associated with KD, doctors may diagnose it as "atypical" KD. Patients presenting with fever and fewer than 4 clinical criteria of KD are labelled as “incomplete” KD. The insufficient phenotype often lead to delayed diagnosis, increasing the risk of coronary complications.

What are the cardiovascular manifestations of KD?

Coronary artery abnormalities (CAA) are observed in approximately 15% to 25% of untreated KD patients. These abnormalities can manifest as widespread dilation (ectasia) and the formation of aneurysms. Myocardial infarction may occur in approximately 2% of all KD patients and in up to 40% of those with persistent aneurysms. Additionally, other cardiac abnormalities may include myocarditis (believed to occur in all KD patients to some extent), valvulitis (typically the mitral valve), pericardial effusion, and aneurysms in systemic arteries.

Treatment of KD

Therapy in acute phase: Intravenous immunoglobulin (IVIG) is very effective when given in the first 10 days of illness. It reduces the chances of development of CAA from 20-25% to 1-2% . The preferred regimen is a single dose of 2 g/kg .

Approximately 10% of KD patients may not respond to IVIG and continue to have persistent fever even 48 hours after administration of the drug(IVIG resistance). These patients may require a second dose of IVIG. Other treatment options to consider in such scenario are corticosteroids and tumor necrosis factor alpha (TNF a) antagonist, infliximab.

Aspirin is administered in patients with KD for anti-inflammatory and antithrombotic effects. During the acute phase, the dose is 70-80 mg/kg/d every 6 hours. After fever subsides, around the 14th day, it is reduced to 3-5 mg/kg/day as a single daily dose which is then continued for several weeks.

Therapy after the acute phase: A follow-up echocardiogram is done 2-3 weeks and again at 6-8 weeks after the illness starts. Aspirin is stopped once inflammatory markers and platelet counts normalise (typically 6-8 weeks) and echocardiograms show no abnormalities. If CAA are found at the 6-8 week echocardiogram, low-dose aspirin should not be discontinued.

The treatment for KD patients with aneurysms depends on the severity of their CAA. Those with extensive or multiple aneurysms may require long-term anticoagulation therapy for management.

Take home message:

1. KD is an important differential in children with exanthematous acute febrile illness.
2. The diagnosis is based on identification of a set of clinical criteria
3. A missed diagnosis of KD can result in severe cardiac complications

COMING UP NEXT - K for Kawasaki Disease….

Dr Suma BALAN

Past Chairperson, PRSI, INDIA (2021-23)

Dr Vijay ViswANAthan

Chairperson PRSI, India (2023-25)

Dr Vighnesh PANdiArajan

Secretary PRSI, India (2023-25)

Dr Himanshi Chaudhary

Consultant Clinical Immunology & Paediatric Rheumatology

Alpha Superspeciality Clinics & Ruby Hall Clinics, Pune

Dr UpendrA KiNjAwadekar

President CIAP (2023)