

## **Kawasaki Disease**

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Kawasaki Disease is an acute, self-limited disease of unknown cause that has a tendency to affect the coronary arteries (the arteries supplying the heart). It predominantly affects infants and children.

Kawasaki Disease is an illness that involves the skin, mouth, and lymph nodes. It is associated with high fevers for 5 or more days.

Kawasaki Disease was first described in Japan in 1967 by Tomisaku Kawasaki; the disease is now known to occur in the Americas, Europe, and Asia in children of all races.

Kawasaki Disease is markedly more common in Japan and children of Japanese ethnicity, having an annual incidence of 150 cases per 100,000 children younger than 5 years of age.

Recurrence rates approach 3%. About 1% has a positive family history. The risk of occurrence in twins is approximately 13%. These data suggest a genetic predisposition may play a role.

The cause of Kawasaki Disease remains a mystery. All the recent evidence suggests an infectious agent, although to date no organism has been found.

This disease occurs in phases. The first phase, which can last for up to 2 weeks, usually involves a persistent fever higher than 104 degrees Fahrenheit and lasts for at least 5 days. The second phase occurs two weeks after the fever begins, and the skin of the hands and feet may begin to peel in large pieces. The child may also experience joint pain, diarrhea, vomiting, or abdominal pain.

Kawasaki disease is a generalized vasculitis (inflammation of blood vessels), affecting all blood vessels throughout the body, but preferentially affects the coronary arteries.

The diagnostic criteria for Kawasaki Disease requires fever for at least 5 days; and four of the five following criteria: 1) Bilateral Conjunctiva injection (blood-shot eyes); 2) Changes of the mucous membranes of the upper respiratory tract: red pharynx, fissured or cracked lips, strawberry appearing tongue; 3) Rash; 4) Changes of the extremities (swelling, redness, skin peeling); 5) Enlarged lymph nodes in the neck region.

A sonogram (Echocardiogram) is done as soon as the diagnosis is entertained to rule out any inflammation of the heart and to check for aneurysm formation of the coronary arteries. It is then done periodically to monitor the condition.

If suspected of having this disease, treatment is started immediately with aspirin, and Intravenous Immunoglobulin. These both have anti-inflammatory effects. The sooner these medications are started, the less likely complications may occur.

If a child is diagnosed with an aneurysm, aspirin is continued indefinitely to prevent rupture of the aneurysm or clot formation.

Coronary artery aneurysms occur in 20-25% of untreated children. Resolution of aneurysms occurs in 1-2 years of approximately 50% of patients. Myocardial Infarction (Heart Attack) caused by a clot in an abnormal coronary artery is the principal cause of death. The greatest risk occurs in the first year after diagnosis.

Recent evidence suggests there may be an incomplete form of Kawasaki Disease occurring in some individuals. Some patients do not fulfill the clinical criteria for Kawasaki Disease and are diagnosed based on echocardiographic findings of coronary artery anomalies. Therefore, strict adherence to the diagnostic criteria may in fact miss some cases. So when should incomplete Kawasaki Disease be considered? If a child has unexplained fever for 5 or more days associated with two or three of the diagnostic criteria mentioned above.

Other diseases that may present in a similar manor include: measles, strep throat, and drug hypersensitivity reactions.

Kawasaki Disease can be quite devastating, and the cause remains a mystery.

It is imperative to seek medical attention as soon as possible, as the earlier it is diagnosed the better the outcome.

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