



KAWASAKI'S DISEASE

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Kawasaki's Disease (KD) is an acute febrile illness of infants and children in which redness of the mucous membranes, skin and tongue are associated with swelling of the hands and feet and subsequent desquamation (peeling of the skin) of the hands and feet. Other signs include swollen lymph nodes of the neck (cervical lymphadenitis), redness and swelling of the eyes (conjunctival congestion), sores in the mouth (stomatitis) and swollen lips (chelitis). After a week or two the skin of the hands and feet begin to peel starting around the nails. Upwards of 20% of patients develop coronary artery complications, however most patients have uneventful recoveries without any long term problems.

The peak age of incidence is 1 year of age with a mean of 2.6 years and it is uncommon over 8 years of age. The cause of KD is not known and is generally seen during the winter and spring of the year. There are 3 phases of the disease.

Phase I: Abrupt onset of fever, lasting around 12 days or so, followed by most of the principal symptoms of the disease condition. This phase develops a red rash usually first seen on the palms and soles that then spreads to involve the torso within a couple days. The most common appearance is a hive-like rash; however it may also resemble measles (morbilliform rash), erythema multiforme or a scarletina like rash. It is more impressive on the hands and feet than the torso and the hands and feet generally develop some swelling as well.

Phase II: This is a subacute phase that can last around 30 days during which fever, arthritis and arthralgia, thrombocytosis, desquamation, and carditis generally resolve. This is the phase with the highest risk of sudden death. The desquamation occurs in this phase with significant peeling of the hands and feet starting at the tips of the digits. In addition most of the dangerous cardiac abnormalities occur such as dysrhythmias, heart failure and left ventricular dysfunction.

Phase III: This is the convalescent period and generally starts 8-10 weeks after the beginning of the illness; it basically starts when all symptoms resolve and lasts until all tests have returned to normal.

The diagnosis of KD depends on several criteria. First there must be an elevated temperature of greater than 102.50 F for at least 5 days; then 4 out of 5 of the following:

- Bilateral conjunctival injection (redness of the eyes).
- At least one swollen lymph node in the anterior cervical lymph nodes (the front of the neck).
- A widespread scarlet fever like erythroderma (redness of the skin), with areas of sharply marginated rash, deeply erythematous maculopapular rash and iris lesions.
- At least one of the following: erythema of the palms and soles, edema of the hands and feet, generalized desquamation around the tips of the fingers and toes.
- At least one of the following changes found in and around the mouth: injected/fissured lips, injected pharynx (back of the mouth) and "strawberry" tongue (a bright red appearance of the tongue).

The majority of people with KD recover without long term complications. However about 20% of patients develop some form of vascular involvement such as: coronary artery aneurysms, myocarditis, heart attack (myocardial infarction), peripheral vascular occlusion, small bowel obstruction or stroke. Historically about 1% of patients die from complications of KD.

Early diagnosis is critical so as to try and prevent cardiovascular complications. Patients should be hospitalized during the phase I period to monitor for complications. The primary treatment is aspirin 100mg/kg/day until the fever has passed, after which the

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dose is reduced to 5-10 mg/kg/day until all lab tests return to normal. High dose intravenous gamma globulin (IVIG) has been used to reduce the risk of coronary aneurysms and myocardial infarction.

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