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Kawasaki Disease

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1. WHAT IS KAWASAKI

1.1 What is it?

This disease was first reported in English medical literature in 1967 by a Japanese paediatrician named Tomisaku Kawasaki (the disease has been named after him); he identified a group of children with fever, skin rash, conjunctivitis (red eyes), enanthem (redness of the throat and mouth), swelling of the hands and feet and enlarged lymph nodes in the neck. Initially, the disease was called "mucocutaneous lymph node syndrome". A few years later, heart complications, such as aneurysms of the coronary arteries (large dilatation of these blood vessels) were reported.
Kawasaki disease (KD) is an acute systemic vasculitis, meaning that there is inflammation of the wall of blood vessels that can evolve to dilatations (aneurysms) of any medium-sized artery in the body, primarily the coronary arteries. However, the majority of children will show only the acute symptoms without cardiac complications.

1.2 How common is it?

KD is a rare disease, but one of the most common vasculitis disorders of childhood, along with Henoch-Schoenlein purpura. Kawasaki disease is described all over the world, although it is much more frequent in Japan. It is almost exclusively an illness of young children. Approximately 85% of children with KD are younger than 5 years, with peak age of incidence at 18–24 months; patients aged less than 3 months or more than 5 years are encountered less commonly, but are at increased risk of coronary artery aneurysms (CAA). It is more common in boys than in

girls. Although cases of KD can be diagnosed any time during the year, some seasonal clustering is known to occur, with an increased number in late winter and spring.

1.3 What are the causes of the disease?

The cause of KD remains unclear, although an infectious origin is suspected to be a triggering event. Hypersensitivity or a disordered immune response, probably triggered by an infectious agent (certain viruses or bacteria), may turn on an inflammatory process leading to inflammation and damage of the blood vessels in certain genetically predisposed individuals.

1.4 Is it inherited? Why does my child have this disease? Can it be prevented? Is it infectious?

KD is not a hereditary disease, although a genetic predisposition is suspected. It is very rare to have more than one member of a family with this disease. It is not infectious and it does not spread from one child to another. At present, there is no known prevention. It is possible, but very rare, to have a second episode of this disease in the same patient.

1.5 What are the main symptoms?

The illness presents with unexplained high fever. The child is usually very irritable. The fever can be accompanied or followed by conjunctival infection (redness of both eyes), without pus or secretions. The child can present different types of skin rash, such as measles or scarlet fever rash, urticaria (hives), papules, etc. The skin rash involves mainly the trunk and the extremities and often the diaper area as well, leading to redness and peeling of skin.

Mouth changes might include bright red cracked lips, red tongue (commonly called "strawberry" tongue) and pharyngeal redness. Hands and feet may also be involved with swelling and redness of the palms and soles. The fingers and toes may appear puffy and swollen. These features are followed by a characteristic peeling of skin around the tip of the fingers and toes (around the second to the third week). More than half of patients will present enlarged lymph nodes in the neck; it is

often a single lymph node of at least 1.5 cm.
Sometimes, other symptoms such as joint pain and/or swollen joints, abdominal pain, diarrhoea, irritability or headaches may be seen. In countries where the BCG vaccine (protection from tuberculosis) is given, younger children may show reddening of the BCG scar area.
The heart involvement is the most serious manifestation of KD, due to the possibility of long-term complications. Heart murmurs, rhythm disturbances, and ultrasound abnormalities may be detected. All the different layers of the heart may show some degree of inflammation, meaning that pericarditis (inflammation of the membrane surrounding the heart), myocarditis (inflammation of the cardiac muscle) and also valve involvement may occur. However, the main feature of this disease is the development of coronary artery aneurysms (CAA).

1.6 Is the disease the same in every child?

The severity of the disease varies from child to child. Not every child has every clinical manifestation and most patients will not develop heart involvement. Aneurysms are seen in only 2 to 6 of 100 children who receive treatment. Some children (especially those younger than 1 year) often show incomplete forms of the disease, meaning that they do not present all the characteristic clinical manifestations, making the diagnosis more difficult. Some of these young children may develop aneurysms. They are diagnosed as atypical KD.

1.7 Is the disease in children different from the disease in adults?

This is a disease of childhood, although there are rare reports of KD in adulthood.