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KAWASAKI DISEASE

This disease was described in 1967 by a Japanese paediatrician called Tomisaku Kawasaki. He recognized a group of children with fever, skin rash, conjunctivitis, enanthem (redness of the throat and oral mucosa), swelling of the hands and feet and enlarged lymph nodes (filters that kill bacteria and viruses) in the neck, initially called mucocutaneous lymph node syndrome. A few years later, heart complications, like aneurysms of the coronary arteries (large dilatation of blood vessels) were reported.

What is it?
Kawasaki disease is an acute systemic vasculitis, meaning there is inflammation of the walls of blood vessels, that can lead to aneurysms, mainly of the coronary arteries (the vessels that supply blood to the heart). Not all children with this disease will develop aneurysms and the majority will have acute symptoms without complications.

How common is it?
Kawasaki disease is a rare disease, but one of the most common vasculitis in childhood, along with Henoch-Schonlein purpura. It is almost exclusively an illness of young children. About 80 of 100 patients are under the age of five. It is slightly more common in boys than girls. Although cases of Kawasaki disease can be diagnosed any time during the year, some seasonal variations can occur with an increased number in late winter and spring. It is much more common among Japanese children, but cases are described all over the world.

What are the causes of the disease?
The cause of Kawasaki disease remains unclear, however, an infectious origin is suspected. Hypersensitivity or a disordered immune response, probably triggered by an infectious agent (certain viruses or bacteria), might turn on an inflammatory process leading to damage of the blood vessels in certain genetically predisposed individuals.

Is it inherited? Why has my child got this disease? Can it be prevented? Is it contagious?
Kawasaki disease is not a hereditary disease, however, a genetic predisposition is suspected. It is very rare to have more than one member of a family with this disease. It is not contagious and cannot be prevented. It's possible, but very rare, to have a second episode of this disease.

What are the main symptoms?
The illness begins with unexplained high fever of at least five days. The child is usually very irritable. The fever can be accompanied, or followed, by conjunctival infection (redness of the eye), without pus, or secretions.

The child can present different types of skin rash, like measles, scarlet fever, urticaria (hives), or papules. The skin rash mainly involves the trunk the extremities and the diaper area.
Mouth changes might include bright red, cracked lips, red tongue, commonly called strawberry tongue and pharyngeal redness.
Hands and feet can also be involved, with swelling and redness of palms and soles. These features are followed (around the second or third week) by a characteristic peeling around the tip of fingers and toes.
More than half the patients will present enlargement of lymph nodes in the neck, it is often just one lymph node of 1.5 cm.
Sometimes, other symptoms, such as joint pain, swollen joints, abdominal pain, diarrhea, irritability and headaches can be seen.
The heart involvement is the most serious manifestation of Kawasaki disease, due to the possibility of long-term complications. Heart murmurs, arrhythmias and ultrasound abnormalities can be detected. The different layers of the heart can show some grade of inflammation, meaning that pericarditis (inflammation of the sheet surrounding the heart), myocarditis (inflammation of the cardiac muscle) and valve involvement can occur. However, the main coronary feature of this disease is the development of coronary aneurysms.

Is the disease the same in every child?
The severity of the disease varies from child to child. Not every patient has every clinical manifestation and most of the patients will not develop heart involvement. Aneurysms are seen in only two of 100 children treated for Kawasaki disease.
Some very young children (under one year of age) 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.

Is the disease in children different from the disease in adults?
This is a disease of childhood, similar forms of this vasculitis can be present in adults but with a different clinical picture.

How is it diagnosed?
A definite diagnosis can be made if unexplained high fever lasts for five or more days plus four of the five following features are present: bilateral conjunctivitis, enlarged lymph nodes, skin rash, mouth and tongue involvement and extremities changes.
If a definite diagnosis is not possible, an incomplete form of this disease should be considered.

What is the importance of tests?
Laboratory findings are not specific for this disease, but reflect the degree of inflammation. Tests include, elevated ESR (usually higher than in other similar illnesses), leukocytosis (increased number of white blood cells), anemia (low count of red blood cells). The numbers of platelets (cells involved in blood clotting) is generally normal in

the first weeks of the disease, but begin to rise in the second week, reaching very high counts.
Patients should undergo periodic examinations and assessment of blood tests until they return to normal.
The electrocardiogram (EKG) and echocardiogram should be obtained initially. The echocardiogram can detect aneurysms by evaluating the shape and size of the coronary arteries. In the case of a child with coronary abnormalities, additional studies and evaluations will be needed.

Can it be treated or cured?
The majority of children with Kawasaki disease can be cured, however, some patients develop heart complications despite the use of proper treatment. The disease cannot be prevented, but the best way to decrease coronary complications is to make an early diagnosis and to start treatment promptly.

What are the treatments?
A child with definite or suspected Kawasaki disease should be admitted to the hospital for observation and monitoring for possible heart involvement.
To diminish heart complications, treatment should be started as soon as the diagnosis is made.
Treatment consists of aspirin and intravenous gammaglobulin, both at high doses. Both treatments will diminish the systemic inflammation, reducing acute symptoms. High dose gammaglobulin are an essential part of treatment as they prevent the occurrence of coronary abnormalities in a high proportion of patients. Corticosteroids may also be prescribed, but less frequently.

What are the side effects of drug therapy?
Gammaglobulin therapy is usually well tolerated. Aspirin treatment can cause gastric intolerance, as well as temporary elevation of liver enzymes.

How long should treatment last for?
The high dose gammaglobulin is given once in the great majority of the patients, but sometimes a second dose may be needed.
A high dose aspirin should be given initially, as long as the fever persists, and then tapered down. The low dose of aspirin is maintained due to its anticoagulant effect on the platelets, this means that the platelets will not stick together and helps to prevent the formation of thrombi (blood clots) inside the aneurysms. Thrombi formation inside the aneurism may lead to cardiac infarction, the most dangerous complication of Kawasaki disease.
A child without coronary abnormalities will receive aspirin for a few weeks, but children with aneurysms should be given it for longer periods.

What about unconventional or complementary therapies?
There is no place for unconventional treatments for this disease.

What kind of periodic check-ups are necessary?

Kawasaki disease patients should have periodic assessments of blood counts and ESR until they return to normal.
Sequential echocardiograms are needed to assess the presence of coronary aneurisms and to follow their course. The frequency by which they should be performed depends on the presence and size of the aneurysm. Most of the aneurysms should resolve.
A pediatric rheumatologist should follow the recovery of these children. In places where a pediatric rheumatologist is not available, the pediatrician, along with the cardiologist, will have to monitor these patients, especially the ones who have had heart involvement.

How long will the disease last for?
Kawasaki disease is an illness with three phases. 1) Acute, which includes the first two weeks when the fever and the other symptoms are present. 2) Subacute, from the second to the fourth week, a period in which the platelet count begins to rise and aneurysms can appear. 3) The recovery phase, from the first to the third month, when all laboratory tests return to normal and some of the blood vessel abnormalities (coronary artery aneurysms) are resolved or diminished in size.

What is the long-term prognosis (predicted course and outcome) of the disease?
For the majority of the patients, the prognosis is excellent, as they will develop a normal life, with normal growth and development.
The prognosis for patients with persistent coronary artery abnormalities depends mainly on the development of stenosis and occlusions (the reduction of the size of the blood vessel due to the formation of blood clots).

Some recommendations for every day life. What about sports? Can the child be vaccinated?
It is recommended not to vaccinate these patients for at least three to six months, as the disease and gammaglobulin treatment affect the immune system and this can last for six months.
Children who did not develop heart involvement will not have any restriction in practising sports or any other daily activity. However, children with coronary aneurysms should consult a paediatric cardiologist regarding participation in competitive activities during adolescence.