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Kawasaki disease

What is Kawasaki disease?

Kawasaki disease is an acute febrile illness with inflammation of small- and medium-sized blood vessels throughout the body, in particular, the coronary arteries (blood vessels around the heart).

It was first described in Japan in 1967 by the paediatrician Dr Tomisaku Kawasaki. It is also known as Mucocutaneous Lymph Node Syndrome.

Kawasaki disease is usually self-limiting and resolves spontaneously without treatment within 4–8 weeks. However, 15–20% of cases will have some damage to coronary arteries and approximately 2% of patients will die from a heart attack.

Who gets Kawasaki disease?

80% of cases occur in children younger than 4 years of age; the peak age is between 1 to 2 years. The disease is very uncommon in children over 14 years old or adults. It is more common in boys than girls.

Although cases of Kawasaki disease have been reported in children of all ethnic origins, the highest incidence remains in children of Asian descent especially Japanese. There are 5000–6000 cases each year in Japan.

What causes Kawasaki disease?

It is most likely caused by a very common infectious organism that only causes problems in a small number of in predisposed individuals. Other theories include:

- Infectious disease caused by bacteria or bacterial superantigens (particularly *Streptococcus pyogenes*) and/or virus: because Kawasaki disease is rarely seen in adults, this suggests that adults may have developed immunity to the causative agent.
- Genetic disposition: individuals of Japanese descent, no matter where they live in the world, are more likely to get the disease. An existing history of the disease within a family makes it more likely another family member will get it.
- Abnormality of the immune system.
- A small possible link with carpet shampoo or living near a body of stagnant water, but neither of these theories have yet been established.

What are the signs and symptoms of Kawasaki disease?

There are several clinically apparent stages of Kawasaki disease.

Acute stage (days 1–11)	<ul style="list-style-type: none"> • Sudden onset of fever that lasts for 1 week or longer and does not respond to antibiotic treatment • An extremely irritable child • Red eyes (bilateral conjunctival injection) • Dry, red and cracked lips with possible bleeding
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	<ul style="list-style-type: none"> • Intensely red and spotty tongue, described as a strawberry tongue • Hands and feet that are red and swollen, so much so that child refuses to walk • Rashes tht involve the trunk, arms and legs, which usually appears within 5 days after onset of fever. The rashes may appear in several forms and can be itchy. Often prominent in the groin. • Enlarged lymph glands • Abnormal liver test • Heart complications in the first stage may include myocarditis and pericarditis
Subacute stage (days 11–21)	<ul style="list-style-type: none"> • Fever, rash and enlarged lymph nodes have usually resolved by this stage. A persistent fever may result in a less favourable outcome becaaaasuse of a greater risk of heart complications • Persistent irritability, poor appetite, and conjunctival injection (red eyes) • Peeling of skin of the fingertips and toes begins • Thrombocytosis (increased number of platelet particles in the blood) may develop with the platelet count topping 1 million (normal range 150–400,000). This increases chance of blood clots • Arthritis and arthralgia (muscle and joint aches) may occur • Heart problems (aneurysms or ballooning of blood vessels) may develop during this stage
Convalescent stage (days 21–60)	<ul style="list-style-type: none"> • Clinical signs begin to disappear and laboratory test results return to normal • Most significant clinical finding that persists through this stage is that coronary artery aneurysms (ballooning of blood vessels to and from the heart) continue to enlarge. This may lead to rupture of the blood vessels, heart attack and death
Late effects/chronic stage	<ul style="list-style-type: none"> • During the first year or two after the illness, coronary aneurysms heal and the amount of coronary artery dilation can become less (amount of healing depends on the amount of damage) • Vessels walls will never return to normal as thickening of these walls occurs during the healing process • Aneurysms formed during an episode of Kawasaki disease is of lifetime significance as these may be the cause of heart disease in adulthood

How is Kawasaki disease diagnosed?

There is no specific lab test that definitively diagnoses Kawasaki disease. The diagnosis is established by meeting the following diagnostic criteria:

Fever persisting at least 5 days

- Generally high and spiking up to 40degC or higher
- No response to antibiotics
- Fever persists for 1 to 2 weeks or longer

In addition to fever, 4 of the following 5 conditions should also be met.

- Changes of hands & feet: swelling and redness followed by peeling of fingertips and toes after fever improves
- Diffuse red rash covering most of the body, usually appearing 5 days after start of the fever (90% of cases)
- Red eyes (bilateral conjunctival injection)
- Mouth changes such as a red tongue (strawberry tongue) and dry cracked lips

- Enlarged lymph glands in the neck, usually unilateral (at least one lymph node with a diameter of 1.5 cm or greater) (50–70% of cases)

Atypical or "incomplete" cases of Kawasaki disease, in which patients have, fever plus four or less of the five principal criteria are becoming more common. In these situations, diagnosis can be confirmed using 2-dimensional echocardiography or coronary angiography (special heart tests) to detect coronary artery disease.

What is the treatment of Kawasaki disease?

Antibiotics are given until a bacterial infection is excluded. Otherwise, initial therapy for Kawasaki disease is aimed at reducing fever and other inflammatory features to help prevent coronary problems.

Treatment should be started within the first 10 days of the onset of illness and include a single dose of intravenous immunoglobulin (purified antibodies collected from many blood donations) and oral aspirin. Immunoglobulin helps to lower the risk of cardiac complications and high dose aspirin helps to reduce high fever experienced in the acute stage. High doses of aspirin should be continued until the patient has a normal temperature for at least several days, at which stage the dose can be reduced for its anti-blood clotting effects. Depending on laboratory evidence of heart vessel abnormalities, aspirin is either stopped or continued indefinitely.

In rare instances long term aspirin therapy in children can be complicated by Reye Syndrome. This rare disorder affecting the liver and the brain is thought to be associated with influenza and [chickenpox](#) infection in conjunction with aspirin therapy. If the child develops influenza or chickenpox, aspirin must be stopped temporarily. Children who need to take aspirin indefinitely should be vaccinated against [chickenpox](#) and influenza. Aspirin therapy also should not be given during the six weeks after a chickenpox vaccine.

What are the complications from Kawasaki disease?

The main complication of Kawasaki disease is development and rupture of coronary artery aneurysms. These aneurysms may also cause heart problems in later life. Other complications include dehydration and limited mobility from joint inflammation.

Related information

On DermNet NZ:

- [Bacterial infections](#)
- [Viral infections](#)

Other websites:

- [Kawasaki disease](#) - emedicine dermatology, the online textbook

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