

# Kawasaki Syndrome

## What Is It?

Kawasaki syndrome, also known as mucocutaneous lymph node syndrome, is a rare illness that typically strikes children younger than age 5. It is a mysterious illness of unknown cause, although some scientists suspect that the cause may be a type of bacterium (such as staphylococcus or streptococcus). Kawasaki syndrome was first identified among Japanese children in 1967, and within nine years, the illness had been reported in American children living in Hawaii. Although researchers assume that the Kawasaki syndrome could have resulted from an infection that was carried somehow between Japan and Hawaii, there has been no confirmation that Kawasaki syndrome can be transmitted via contact between people. Recent evidence suggests that inherited (genetic) factors may also be important, but the cause of Kawasaki syndrome remains a mystery.

Kawasaki syndrome makes children ill by triggering inflammation in many different parts of the body. In the heart, this inflammation can take the form of **myocarditis** (inflammation of heart muscle), **pericarditis** (inflammation of membranes covering the heart) or valvulitis (inflammation of the heart valves). Kawasaki syndrome also can cause a type of **meningitis** (inflammation of membranes covering the brain and spinal cord), and it can produce inflammation in the skin, eyes, lungs, lymph nodes, joints and mouth. However, the most important problem related to Kawasaki syndrome is the threat of **vasculitis** (blood-vessel inflammation), especially in the body's medium-sized arteries. This vasculitis can be especially dangerous when it damages the heart's coronary arteries, producing an abnormal widening (dilation) or aneurysm (bulge) in these vessels. In rare cases, artery damage related to Kawasaki syndrome can significantly interfere with the heart's blood supply, even to the point of causing a **heart attack** in a very young child.

In the United States and other industrialized nations, Kawasaki syndrome is now the most common cause of heart disease that is not present at birth in children. According to U.S. government statistics, a total of 1,262 cases of Kawasaki syndrome were reported between 1994 and 1997, with an average of 315 cases each year. Approximately 80 percent of these patients were younger than 5 years old, and boys were affected more often than girls.

## Symptoms

Because there is no test available to confirm that a person has Kawasaki syndrome, doctors define the condition by the symptoms that it produces. The most important symptom is a high fever (usually 104 degrees Fahrenheit or above) for at least five days. This fever must occur together with at least four of the following additional symptoms:

- Conjunctivitis** of both eyes (bloodshot eyes)

- Symptoms involving the mouth or throat, including redness and inflammation of the lips or throat, cracked lips, bleeding lips or a strawberry-colored tongue

- Symptoms affecting the hands or feet, including swelling, redness of the skin on the palms and soles or peeling skin on the fingertips, toes, palms, or soles

- A rash, primarily on the torso

Swollen glands in the neck

People with Kawasaki syndrome can have other symptoms that are not part of the definition of the disease. These can include:

Pain and swelling of the joints

Diarrhea

Vomiting

Abdominal pain

Cough

Earache

Runny nose

Irritability

Seizures

Weakness in the arms or legs

Weakness of facial muscles

Heart-rhythm abnormalities

Signs of heart failure

## **Diagnosis**

Because Kawasaki syndrome is rare in the United States, your doctor will want to rule out other illnesses that are more common and produce similar symptoms.

If your doctor suspects that your child has Kawasaki syndrome, he or she may begin by asking you about:

Your child's medications to rule out a drug reaction

Any recent exposure to someone with strep throat to rule out scarlet fever, which is caused by a streptococcal infection

Any recent exposure to someone with measles to rule out measles if your child has not been immunized against this illness

Any recent tick bite to rule out Rocky Mountain spotted fever

As part of the diagnostic work-up for Kawasaki syndrome, your doctor may need to order blood tests and special diagnostic procedures to rule out other infectious or noninfectious causes of your child's symptoms. Once it becomes clear that Kawasaki syndrome is a possibility, your doctor may schedule an echocardiogram, a painless test that uses sound waves to outline the heart's structure, to check for any dilation or aneurysms in the coronary arteries. In some cases, your doctor also may recommend coronary angiography, a procedure in which dye is injected into the arteries that supply the heart to look for evidence of aneurysm or narrowing typical of Kawasaki syndrome.

Your doctor will diagnose Kawasaki syndrome if your child has symptoms that fit the definition of the illness and if there is no other explanation for these symptoms. Researchers continue to look for a reliable diagnostic test for this illness, but, so far, none is available.

## **Expected Duration**

Kawasaki syndrome lasts for several weeks, progressing through three different stages:

**Acute phase** — This is the most intense part of the illness, when symptoms are most severe. It usually lasts one to two weeks.

**Subacute phase** — This stage begins when the child's fever, rash and swollen lymph nodes go away. However, the child still feels irritable, has a poor appetite and slight eye redness and may develop peeling skin on the fingers and toes. This stage usually ends three to four weeks after the fever began.

**Convalescent stage** — This stage begins when all clinical symptoms are gone, but the results of a blood test called the erythrocyte sedimentation rate (ESR), an indicator that there is still inflammation in the body, remain abnormal. When the ESR results finally return to normal, usually six to eight weeks after the fever began, the convalescent stage ends.

## **Prevention**

Because the cause of Kawasaki syndrome is unknown, there is no known way to prevent it.

## **Treatment**

Kawasaki syndrome is treated with a single dose of gamma globulin, given intravenously (directly into a vein), together with aspirin taken by mouth. Gamma globulin is a purified collection of proteins and antibodies from donated blood. It is not known why it is effective against this disease. The gamma-globulin injections may be repeated over several days if fever persists.

Parents generally are warned against giving aspirin to children because it can trigger Reye's syndrome, a serious disorder that can damage the brain and liver. However, aspirin is the best treatment for Kawasaki syndrome, so the benefits outweigh the risk of Reye's syndrome, which is rare. The child usually continues to take aspirin for six to eight weeks after the acute symptoms of Kawasaki syndrome subside.

Other treatments, including corticosteroids or immunosuppressive medications are not always helpful, but sometimes are recommended if gamma globulin injections are not effective. A procedure called plasmapheresis sometimes is recommended for Kawasaki syndrome that does not respond to other treatments. In plasmapheresis, blood is removed, filtered to remove proteins (including antibodies) and then returned to the body.

## **When To Call A Professional**

Call your doctor immediately if your child develops a high fever, with or without other symptoms of Kawasaki syndrome.

## **Prognosis**

Without proper treatment, 20 percent to 25 percent of patients with Kawasaki syndrome develop abnormalities in their coronary arteries. With treatment, this percentage drops to 2 percent to 4 percent. In general, the risk of death from cardiac complications is about 1 percent to 2 percent, most commonly during the first two to 12 weeks of illness.

Up to 25 percent of patients with Kawasaki syndrome have coronary aneurysms during the first one to three months of illness. The aneurysms often go away on their own within the next year or two. Children

who continue to have coronary abnormalities usually need regular testing, including stress tests, echocardiography and sometimes coronary angiography. If cardiac health deteriorates because of chronic damage to the coronary arteries, surgery may be needed to correct the problem.

Recent Japanese studies have shown that the death rate for people with Kawasaki syndrome is about the same after the acute phase has passed as for people without Kawasaki syndrome.

### **Additional Info**

Office of Rare Diseases  
National Institutes of Health  
31 Center Dr., MSC 2084  
Building 31, Room 1B-19  
Bethesda, MD 20892-2084  
Phone: (301) 402-4336  
Fax: (301) 480-9655  
E-Mail: [hh70f@nih.gov](mailto:hh70f@nih.gov)  
<http://rarediseases.info.nih.gov/>

National Institute of Allergy and Infectious Diseases (NIAID)  
Building 31, Room 7A-50  
31 Center Dr. MSC 2520  
Bethesda, MD 20892-2520  
Phone: (301) 496-5717  
<http://www.niaid.nih.gov/>

National Center for Infectious Diseases  
Traveler's Health  
Centers for Disease Control and Prevention  
1600 Clifton Rd., NE  
Atlanta, GA 30333  
Toll-Free: (800) 311-3435  
Fax: (888) 232-3299  
<http://www.cdc.gov/travel/>

National Heart, Lung and Blood Institute (NHLBI)  
6701 Rockledge Dr.  
P.O. Box 30105  
Bethesda, MD 20824-0105  
Phone: (301) 592-8573  
E-Mail: [NHLBIinfo@rover.nhlbi.nih.gov](mailto:NHLBIinfo@rover.nhlbi.nih.gov)  
<http://www.nhlbi.nih.gov/>