

What is Kawasaki Disease?

Kawasaki disease is a form of vasculitis. Vasculitis is a group of rare disorders characterized by inflammation of the blood vessels. Inflammation of blood vessels may result in aneurysms or narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues.

Kawasaki disease is also called mucocutaneous lymph node syndrome, and it affects the mucus membranes, lymph nodes, and the arteries in the heart.

Vasculitis is classified as an autoimmune disorder—a disease that occurs when the body's natural defense system mistakenly attacks healthy tissues. The inflammatory process in vasculitis may be triggered by genetic or environmental factors, drug or vaccine reactions, or an infection or virus.

Causes

The cause of Kawasaki disease is not fully understood. It appears that children of parents who have had Kawasaki disease are twice as likely to develop the it compared to the general population, but the genetic link is still not well understood. The disease tends to occur in outbreaks, indicating a virus may play a role in Kawasaki disease.

Who Gets Kawasaki Disease?

Kawasaki disease is considered rare, and one that almost always affects children under the age of 5. In very rare cases, the disease may occur during adolescence or adulthood. Boys are almost twice as likely to develop the disease as girls.

The disease is reported around the world, but the highest incidence is in Japan. It can occur in any racial or ethnic group, but children of Asian or Pacific Island descent have higher rates of the disease. Estimates indicate approximately 4,000 cases of Kawasaki disease are diagnosed each year in the United States, most frequently among children of Asian-American background. The disease occurs year-round but is mostly seen in winter and early spring.

Symptoms

The first signs of Kawasaki disease are:

- High fever that lasts five days or more (101-104°F and above)
- Irritability

Your child may also have the following symptoms:

- Swollen mouth, nose, and throat (including red cracked lips and a red swollen tongue)
- Swollen lymph glands in the neck
- Bloodshot eyes
- A rash on the trunk of the body and groin
- Swollen hands and feet
- Peeling skin on the hands and feet, especially the tips of the fingers and toes
- Joint pain
- Stomach pain, diarrhea, and vomiting

If your child has a fever or other symptoms as noted above, report them to the doctor as soon as possible. Regular check-ups and ongoing monitoring of lab and imaging tests are important in managing the condition.

Early diagnosis and treatment are important to prevent heart damage.

Diagnosis

There is no single test for diagnosing Kawasaki disease, so your child's doctor will consider several factors, including a detailed medical history and physical examination, as well as:

- Laboratory tests, including blood and urine tests
- Specialized imaging studies, including x-rays and computed tomographic scan
- A biopsy of an affected tissue or organ, when indicated

These will help to rule out other causes of fever or diseases with similar symptoms, such as measles, scarlet fever, Rocky Mountain spotted fever, and juvenile rheumatoid arthritis.

To detect heart problems your doctor may perform an:

- Echocardiogram (a cardiac ultrasound), which checks the heart valves and function
- Electrocardiogram, which records the electrical activities of the heart muscle.

Echocardiograms or other diagnostic tests may need to be repeated at certain intervals to monitor for heart damage following recovery from this disease.

Treatment

Once diagnosed, early treatment is very important in reducing symptoms and minimizing heart damage, ideally within the first 10 days. The standard treatment, which is given in the hospital includes:

- Intravenous immunoglobulin therapy (IVIG), a human blood protein that helps prevent damage to the heart blood vessels
- Aspirin to reduce fever, rash, joint swelling, and pain
 - **(Note:** Never give aspirin to your child without a doctor's orders. Aspirin use in children and teenagers has been associated with Reye's syndrome, a rare but serious condition that causes swelling in the brain and liver.)

If IVIG does not work corticosteroids or medications that suppress the immune may be given.

Rarely, surgery may be required to repair damaged heart blood vessels.

Children diagnosed with Kawasaki disease will likely require follow-up medical care, particularly if the disease has affected the heart.

Side Effects

The medications used to treat Kawasaki disease have potentially serious side effects, such as:

- Lowering the body's ability to fight infection

It is important to monitor your child's symptoms and report them to the doctor.

Infection prevention is also very important. Talk to the doctor about age-appropriate vaccinations, which can reduce your child's risk of infection.

Relapse

Even with effective treatment, relapses can occur with Kawasaki disease. If your child's fever or other symptoms return, report them to the doctor as soon as possible.

Regular check-ups and ongoing monitoring of lab and imaging tests are important in detecting relapses.

Your Child's Medical Team

Effective treatment of Kawasaki disease may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a pediatrician or primary care doctor, your child may need to see the following:

- Pediatric cardiologist (specialist in diagnosis and treatment of heart disorders in children)
- Pediatric or adult infectious disease specialist (to help rule out infectious disease as a cause of fever)
- Pediatric or adult rheumatologist (joints, connective tissue, and immune system)
- Pediatric dermatologist (skin)
- Others as needed

The best way to manage Kawasaki disease is to actively partner with your child's health care providers and get to know the members of your health care team.

It may be helpful to keep a health care journal to track your child's medications, symptoms, test results, and notes from doctor appointments in one place.

To get the most out of doctor visits, make a list of questions beforehand and bring along a supportive friend or family member, if necessary, to provide a second set of ears and take notes.

Remember, it's up to you to be your child's advocate. If you have concerns with the treatment plan, be sure to speak with the medical team. It's always your right to seek a second opinion.

Outlook

Most children with Kawasaki disease experience a full recovery. However, 1 in 4 may develop serious heart problems, so early diagnosis and treatment are critical in preventing heart damage. A very small percentage of patients die of complications from coronary blood vessel inflammation.

Children with Kawasaki disease should have follow-up echocardiograms and checkups to screen for ongoing heart problems.

(Revised October 2020)