

## KAWASAKI DISEASE

### What is Kawasaki disease?

Kawasaki disease is a form of vasculitis—a family of rare disorders characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Kawasaki primarily occurs in children from 6 months to age 5. Also called mucocutaneous lymph node syndrome, the disease affects the mucus membranes, lymph nodes, and the coronary arteries which supply blood to the heart muscle. It is the leading cause of acquired heart disease in children in the United States.

Common symptoms include fever, rash, swollen lymph glands in the neck, inflammation of the mouth, nose and throat, eye irritation and redness, swelling of the hands and feet, and peeling skin. On their own, these symptoms are rarely serious, and most children recover within a few weeks. However, 1 in 4 children with Kawasaki may develop serious heart problems.

Once diagnosed, early treatment is vital in reducing symptoms and minimizing heart damage. The standard treatment for Kawasaki includes intravenous immunoglobulin therapy (IVIG), plus aspirin, administered in a hospital. Most individuals respond well to IVIG, but some require corticosteroids or medications that suppress the immune system. Children diagnosed with Kawasaki will likely require follow-up medical care, particularly if the disease has affected the heart.

### Causes

The cause of Kawasaki disease is not fully understood by researchers. Vasculitis is classified as an autoimmune disorder—a disease which 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. It appears that children of parents who have had Kawasaki are twice as likely to develop the disease 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.

### Who gets Kawasaki disease?

Kawasaki is considered a rare disease 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, mostly 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 usually a persistent high fever and irritability. The fever can range from moderate (101-103°F), to high (104°F and above), and lasts five days or more. Your child may also have the following symptoms:

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- 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

The most serious complication is heart involvement caused by inflammation of the coronary arteries that bring blood to the heart muscle. This can result in aneurysms (an abnormal bulging of the arteries which can rupture), inflammation of the heart muscle, heart valve problems or, very rarely, heart attack.

### Diagnosis

There is no single test for diagnosing Kawasaki, so your child's doctor will consider a number of factors, including a detailed medical history; physical examination; laboratory tests; specialized imaging studies; and, when indicated, a biopsy of an affected tissue or organ.

The presence of classic symptoms help point to a diagnosis of Kawasaki, including fever lasting at least five days and at least four of five of the following: swollen lymph nodes; bloodshot eyes; swollen lips/mouth/tongue; swollen hands and feet; and rash. The doctor will try 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.

If Kawasaki disease is suspected, your child's doctor will likely order blood and urine tests, as well as chest X-rays to look for changes or damage to the aorta. Common diagnostic tests to detect potential heart involvement include the echocardiogram (a cardiac ultrasound), which checks the heart valves and function; and electrocardiogram (EKG), 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

To prevent complications, treatment should begin as soon as possible after diagnosis, ideally within the first 10 days. Most children with Kawasaki require treatment in a hospital. Standard treatment includes high doses of IVIG, a human blood protein that helps prevent damage to the heart blood vessels, if given early in the disease. Aspirin is given 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.)

Patients usually improve significantly within 24 hours of treatment with IVIG. However, some patients may need to take corticosteroids or immunosuppressant medications if initial treatments aren't working. Rarely, surgery may be required to repair damaged heart blood vessels.

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### Side effects

The medications used to treat Kawasaki have potentially serious side effects, such as lowering the body's ability to fight infection, among others, so 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. 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 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); and others as needed.

The best way to manage Kawasaki disease is to actively partner with your child's health care providers. 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

The majority of children with Kawasaki 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. Patients with Kawasaki should have follow-up echocardiograms and checkups to screen for ongoing heart problems.

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC), to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information.

For more information on vasculitis research, visit: [www.vasculitisfoundation.org/research](http://www.vasculitisfoundation.org/research)

## **KAWASAKI DISEASE**

### **About Vasculitis**

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

### **A Family of Diseases**

- Anti-GBM (Goodpasture's) disease
- Aortitis
- Behcet's syndrome
- Central nervous system vasculitis
- Cogan's syndrome
- Cryoglobulinemia
- Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (Henoch-Schönlein Purpura)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Rheumatoid vasculitis
- Takayasu's arteritis
- Urticarial vasculitis

### **About the Vasculitis Foundation**

The Vasculitis Foundation (VF) is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services. To learn more, and get the most updated disease and treatment information, visit [www.vasculitisfoundation.org](http://www.vasculitisfoundation.org)

### **Connect with the VF on Social Media**

- Instagram: [vasculitisfoundation](https://www.instagram.com/vasculitisfoundation)
- Twitter: [@VasculitisFound](https://twitter.com/VasculitisFound)
- VF Facebook Discussion Group: [www.facebook.com/groups/vasculitisfoundation](https://www.facebook.com/groups/vasculitisfoundation)

## **KAWASAKI DISEASE**

### **Ways to Get Involved**

- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium
- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

### **Vasculitis Foundation Mission**

Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.

### **Join the VPPRN!**

The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: [www.VPPRN.org](http://www.VPPRN.org)

The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: [www.rarediseasesnetwork.org/cms/vcrc](http://www.rarediseasesnetwork.org/cms/vcrc)

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