

# Kawasaki Disease

Kawasaki disease is a condition involving inflammation of the blood vessels in young children. Children with this disease develop a high fever lasting five or more days, rash, red eyes and lips, and other symptoms. The main danger is a risk of damage to the blood vessels in the heart. Treatment is needed to reduce the chances of this complication. With treatment, most children with Kawasaki disease recover completely.

## What is Kawasaki disease?

Kawasaki disease is a potentially serious disease involving inflammation of the blood vessels (vasculitis) in children, mainly under age 5. The cause is unknown. Children with Kawasaki disease can become very ill for a week or two, then take a long time to recover if untreated.

Although most children recover completely, there is a risk of damage to the coronary arteries supplying blood to the heart. This complication occurs in about 20% of children with Kawasaki disease if not treated. With appropriate treatment, the risk drops to 5%. The longer the initial fever lasts, the higher the risk.

Children with Kawasaki disease need timely treatment and close follow-up during the recovery period. If there are no complications, your child should recover completely over a period of several weeks.

## What does it look like?

The first symptom of Kawasaki disease is fever:

- Fever may be very high—up to 104°F (40°C).
- Fever comes and goes.
- It usually lasts 1 to 2 weeks, if untreated.
- It does not improve with antibiotics.

Other typical symptoms are:

- Red eyes.
- Red, dry, cracked lips; red, swollen mouth and throat, including “strawberry tongue.”
- Redness and swelling of the palms and soles, followed by peeling of the skin on the fingers and toes.
- Various skin rashes.
- Swelling of the lymph nodes in the neck, usually on one side.
- During the illness, children are usually very fussy. Other, less common symptoms include:

- Joint swelling.
- Vomiting.
- Inflammation of the urethra (opening where urine comes out).
- Swollen gallbladder.

The early, acute phase of Kawasaki disease lasts 1 to 2 weeks. Illness becomes less severe for about a month afterward. The risk of coronary artery disease is highest during this period. Your child then slowly gets better over another month or two. All symptoms are much shortened by appropriate treatment.

*Heart involvement* is the most serious problem. Various parts of the heart may be affected. However, permanent damage can result from inflammation of the coronary arteries, which supply blood to the heart muscle. 

## What causes Kawasaki disease?

The exact cause is unknown.

## What are some possible complications of Kawasaki disease?

Heart problems are the main complication of Kawasaki disease. Most of the symptoms result from inflammation of the blood vessels.

- *Myocarditis or pericarditis.* Inflammation of the heart muscle or heart lining. Early in the illness, these complications may cause fast heartbeat or reduced heart function.
- *Coronary artery aneurysms.* Inflammation of the vessels that supply blood to the heart muscle can lead to “weak spots” in the vessel walls, causing them to balloon out or dilate. These weak spots, called aneurysms, can develop a blood clot (thrombosis). The clot can prevent enough blood from getting to the heart muscle, causing problems such as myocardial infarction (MI, or “heart attack”).
- *Without treatment,* about one fifth of children with Kawasaki disease develop aneurysms. The risk is highest in the second or third week of illness.
- *With treatment,* the risk of coronary artery damage is reduced to less than 5%.

As long as heart and blood vessel damage does not occur, most children with Kawasaki disease recover completely. Even without treatment, about 50% of aneurysms go away. There may still be some mild abnormalities, such as narrowing of the blood vessels.

## What puts your child at risk of Kawasaki disease?

- Kawasaki disease almost always occurs in infants and children under age 5.
- The risk is highest in Asian children, but any racial/ethnic group can be affected.

## Can Kawasaki disease be prevented?

There is no known way to prevent Kawasaki disease.

## How is Kawasaki disease diagnosed?

There is no specific test for Kawasaki disease. The diagnosis is based on symptoms, with the help of some blood tests, especially tests for inflammation such as the erythrocyte sedimentation rate (ESR). Increased platelet counts (blood particles that help stop bleeding) may be present.

Because of the risk of heart and blood vessel complications, it's important to diagnose Kawasaki disease promptly. Unfortunately, Kawasaki disease can be difficult to recognize; at first, it may be confused with viral infections, strep infections, or others. Diagnosis is especially difficult if your child doesn't have the typical symptoms.

Once the disease is recognized, a test called echocardiography ("echo") may be done to check for damage to the heart and blood vessels. This painless test uses sound waves to take pictures of your child's heart. It may be repeated a few times to look for aneurysms and other abnormalities. A specialist in children's heart diseases (a pediatric cardiologist) will supervise this part of your child's care.

## How is Kawasaki disease treated?

Once Kawasaki disease is recognized, your child will be admitted to the hospital for treatment.

- Treatment consists of giving *immune globulin*, which is the part of our blood that contains our antibodies. When given intravenously it is called IVIG. Antibodies are made by our immune system to fight infection. *Aspirin*

is also given, at first for inflammation and then to prevent any clots from forming on aneurysms that might be present.

- For most children, this treatment quickly reduces fever and other symptoms.
- Treatment greatly reduces the risk of heart and blood vessel complications.
- Treatment is most effective if started within 10 days after the start of your child's illness. Treatment is usually still given if Kawasaki disease is recognized later on, but it is not known how effective it is.
- Your child's doctors will closely monitor his or her condition during treatment. If the response is not as good as expected, treatment may be repeated or other drugs (such as steroids tried).
- If your child's tests find no heart or blood vessel abnormalities, aspirin treatment should continue at a lower dose for 6 to 8 weeks from the start of illness. It may take several more weeks before your child recovers completely.
- If heart or blood vessel abnormalities are present, your child will need further treatment. Echocardiograms will be done to check on your child's condition.
- Medications such as aspirin and warfarin (coumadin) are used to prevent clotting. Blood clots can be removed if necessary.
- Small aneurysms usually clear up in a year or two, but some abnormalities may remain. If your child has a small aneurysm, he or she may have to continue taking aspirin for a long time.
- If your child has a large aneurysm or more than one aneurysm, other treatments may be needed.
- With close medical follow-up, the risk of death from complications of Kawasaki disease is low.

## When should I call your office?

Call the pediatric cardiologist, or our office, if you have questions regarding Kawasaki disease and your child's treatment and recovery.