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Kawasaki Disease

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Kawasaki disease is an acute childhood illness that is characterized by inflammation of the blood vessels. Early diagnosis and treatment are important to prevent possible damage to the coronary arteries, which supply oxygen to the heart muscle.

The classic symptoms of Kawasaki disease are listed below:

- Fever lasting for at least 4 days
- Red eyes (conjunctivitis), usually without drainage
- Redness of the lips, throat, and tongue (strawberry tongue)
- Redness and swelling of the hands and feet, followed later by peeling of skin on the fingers and toes
- A rash
- Swollen gland in the neck (usually 1 large) measuring at least 1.5 cm or larger

Although children with Kawasaki disease usually have fever plus 4 of the 5 signs listed above, some children have fewer symptoms (incomplete Kawasaki disease). If your child has a fever for 5 or more days with 2 or 3 of these classic symptoms, without another diagnosis, you should bring your child to the doctor or other healthcare provider. In addition to the signs listed

above, children with Kawasaki disease are often very fussy or irritable. Vomiting, diarrhea, abdominal pain, respiratory symptoms such as cough, and joint pains are common in the days before diagnosis.

What Heart Problems Can Occur in Kawasaki Disease?

Inflammation in the walls of the coronary arteries can cause them to weaken. The weakened artery forms an aneurysm, which is a bubble-shaped (saccular) or cigar-shaped (fusiform) enlargement (Figure). Without timely treatment, 1 in 5 children with Kawasaki disease will develop coronary aneurysms. Treatment in the first 10 days of illness reduces the risk of aneurysm formation to less than 5%. Kawasaki disease can also cause inflammation of the heart muscle (myocarditis) and leakiness of heart valves, but these problems are almost always temporary.

What Causes Kawasaki Disease?

The cause of Kawasaki disease is unknown. Kawasaki disease is not spread from person to person. Many experts think that Kawasaki disease

is an immune reaction triggered by an infection.

Who Is at Risk?

Although the risk is highest in those of Japanese origin, children of all races and ethnic groups can develop Kawasaki disease. Most children are younger than age 5 years, but even older children and teenagers can be affected. The risk of developing coronary artery aneurysms is greatest for children (1) who are younger than age 6 months, (2) whose diagnosis and treatment are delayed, or (3) who continue to have fever despite treatment.

What Is the Typical Course of Kawasaki Disease?

The symptoms of Kawasaki disease usually resolve within 4 to 6 weeks, but return of your child's usual personality and energy level may take as long as 8 weeks.

How Is Kawasaki Disease Treated?

Kawasaki disease is treated with intravenous immunoglobulin (IVIG), given through an intravenous line in the hospital over 8 to 12 hours. For most patients, IVIG improves fever and

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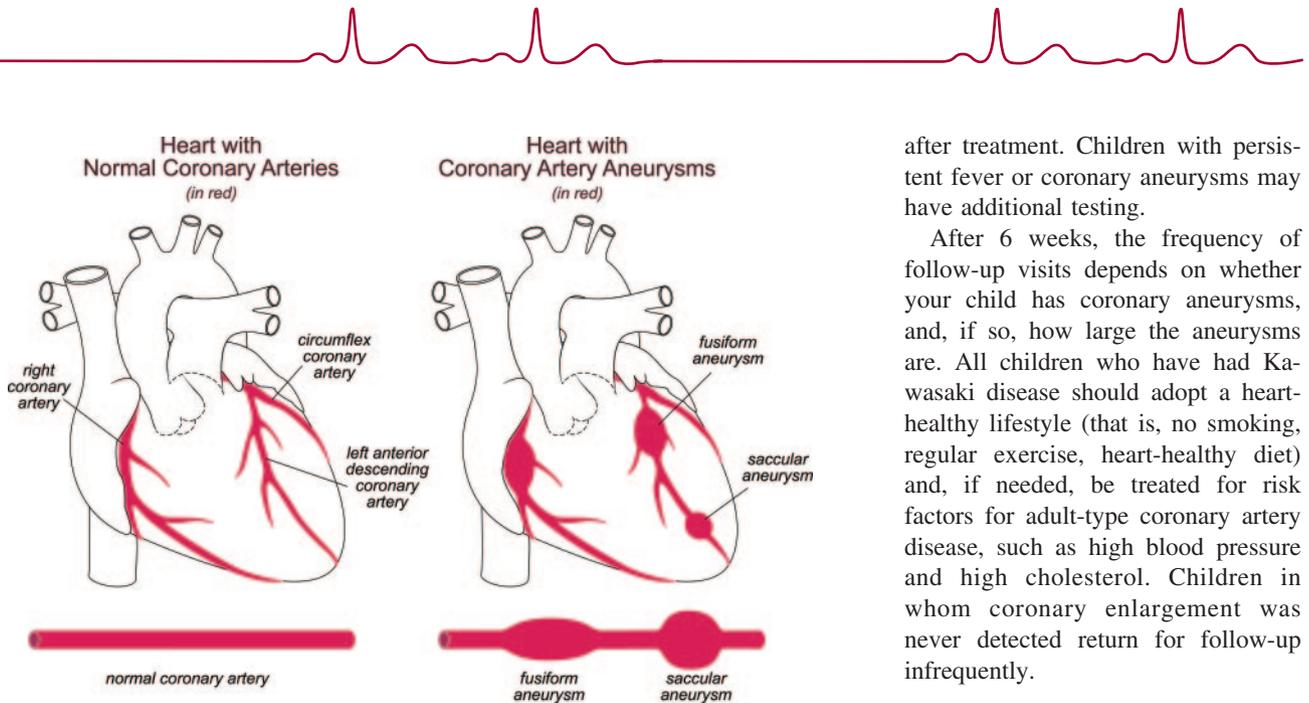


Figure. Coronary arteries are demonstrated in red. The panel on the left shows a heart with normal coronary arteries, and the panel on the right shows a heart with large aneurysms of 2 types: fusiform (cigar shaped) and saccular (bubble shaped).

lowers the risk of coronary aneurysms if administered in the first 10 days, and ideally within 7 days, of the start of fever. Aspirin also is prescribed in high doses to control fever. Once the fever is gone, the aspirin dose is lowered to a single daily dose to prevent blood from clotting in the coronary arteries. If no aneurysms are present by 5 or 6 weeks after the onset of fever, aspirin is stopped. Children with persistent fever or worsening aneurysms despite IVIG therapy are often treated with more IVIG infusions and/or other therapies to lessen inflammation. For children who have developed coronary aneurysms, treatment is aimed at preventing blood clots in the coronary arteries. Aneurysms usually reach their biggest size about 4 weeks after the onset of fever. Depending on the size of the aneurysms, children may be treated with at least 1 of the following medications: low-dose aspirin, clopidogrel (Plavix), warfarin (Coumadin), or low-molecular-weight heparin (Lovenox). Such treatments are continued long-term. Some patients with very large aneurysms or those who have had heart attacks may need additional medicines.

What Happens to Coronary Aneurysms?

Half of aneurysms heal to a normal size after 1 to 2 years, although healing may cause the wall of the artery to become thicker than usual. The smaller the initial size of the aneurysm, the more likely it is to heal to its normal size.

Over years, blockage (stenoses) may develop in damaged coronary arteries, usually at either end of an aneurysm. Children with the largest aneurysms are the most likely to develop coronary artery stenoses. If the heart muscle does not receive enough oxygen (for example, because blood flow through a coronary artery is impaired by stenosis or by a blood clot), heart pain (angina) or a heart attack may occur. New blood vessels (collaterals) often grow to supplement the blood supply to the heart muscle territory serviced by a gradually blocking coronary artery.

How Are Children Followed Up After Kawasaki Disease?

In the first 6 weeks of the illness, children usually have blood tests and an echocardiogram on a minimum of 3 occasions: at the time of diagnosis and then 1 to 2 weeks and 4 to 6 weeks

after treatment. Children with persistent fever or coronary aneurysms may have additional testing.

After 6 weeks, the frequency of follow-up visits depends on whether your child has coronary aneurysms, and, if so, how large the aneurysms are. All children who have had Kawasaki disease should adopt a heart-healthy lifestyle (that is, no smoking, regular exercise, heart-healthy diet) and, if needed, be treated for risk factors for adult-type coronary artery disease, such as high blood pressure and high cholesterol. Children in whom coronary enlargement was never detected return for follow-up infrequently.

Follow-Up Testing for Children With Aneurysms

The types of testing for children with aneurysms are as follows:

Electrocardiogram: Records electrical activity in the heart.

Echocardiogram: Ultrasound of the heart to image coronary arteries, as well as heart muscle and heart valve function.

Cardiac magnetic resonance imaging or computed tomography angiography: Shows shape and size of the coronary arteries when they cannot be well seen by echocardiography (for example, in older children).

Stress tests with myocardial imaging: Depending on the age of your child, stress testing may be done by use of a medication (babies and children under age 5 years) or by exercise (in older children). The response of the heart to stress may be assessed by echocardiogram, cardiac magnetic resonance imaging, or nuclear imaging.

Heart catheterization (coronary angiography): A tube is threaded through a leg artery to the heart, and pictures are taken of the coronary arteries to show their size and shape.

What Can Be Done if a Coronary Blockage Is Found?

If coronary artery blockage is found at cardiac catheterization, it may be opened with use of a balloon or a tube

placed inside the vessel (stent). Coronary blockages in some patients are better treated with coronary artery bypass surgery. The majority of children return to normal lives after bypass graft surgery for Kawasaki disease.

Summary

Kawasaki disease is an illness with potentially serious effects on the heart. Current therapy is focused on the early treatment and close monitoring of cardiac status. Children who never developed aneurysms (that is, whose coronary arteries were always normal) have a rate of heart problems similar to that found in the general population; research studies are exploring whether arteries in these children might be

slightly stiffer or thicker than usual. Most children with coronary aneurysms take part in normal activities but need to see their cardiologist regularly. These children/adults may have some activity restrictions depending on the size of their aneurysms and whether or not they are on medicines that slow blood clotting. All children who have had Kawasaki disease should maintain a heart-healthy lifestyle and reduce risk factors for hardening of the arteries (atherosclerosis), such as high cholesterol, high blood pressure, or smoking.

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Additional Resources

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