



# Kawasaki Disease Evaluation and Care



**NATIONWIDE  
CHILDREN'S®**

*When your child needs a hospital, everything matters.*

## What is Kawasaki disease?

Kawasaki disease (KD) is a rare condition in children that involves inflammation of the blood vessels. Symptoms often begin with high-grade fever that lasts for at least five days and is unresponsive to over-the-counter medication. Kawasaki disease represents the most common cause of acquired heart disease in children in the developed world, as the inflammation can affect a patient's coronary arteries and heart.

## Recognizing Kawasaki Disease

While Kawasaki disease can't be prevented, it usually has telltale symptoms and signs that appear in phases. The first phase, which can last for up to two weeks, usually involves a persistent fever higher than 104°F (39°C) that lasts for at least five days. During the second phase, which usually begins within two weeks of the onset of fever, the skin on the hands and feet may begin to peel in large pieces. The child also may experience joint pain, diarrhea, vomiting or abdominal pain.

### Signs and Symptoms

- Fever that lasts for several days
- Rash (often worse in the groin area)
- Red eyes without drainage or crusting
- Bright red, swollen, cracked lips
- Strawberry tongue appearing with shiny, bright red spots after the top coating sloughs off
- Swollen hands and feet
- Redness of the palms and soles of the feet
- Swollen lymph nodes
- Peeling of fingertips and toes
- Temporary arthritis, enlarged gall bladder, temporary hearing loss, abdominal pain, vomiting and diarrhea

## Potential Complications

Physicians can manage the symptoms of Kawasaki disease if they catch it early. Symptoms often disappear within just two days of the start of treatment. If Kawasaki disease is treated within 10 days of the onset of symptoms, heart problems usually do not develop.

Cases that go untreated can lead to more serious complications, such as vasculitis, an inflammation of the blood vessels. This can be particularly dangerous because it can affect the coronary arteries, which supply blood to the heart. In addition to the coronary arteries, the heart muscle, lining, valves and the outer membrane that surrounds the heart can become inflamed. Arrhythmias (changes in the normal pattern of the heartbeat) or abnormal functioning of some heart valves also can occur.

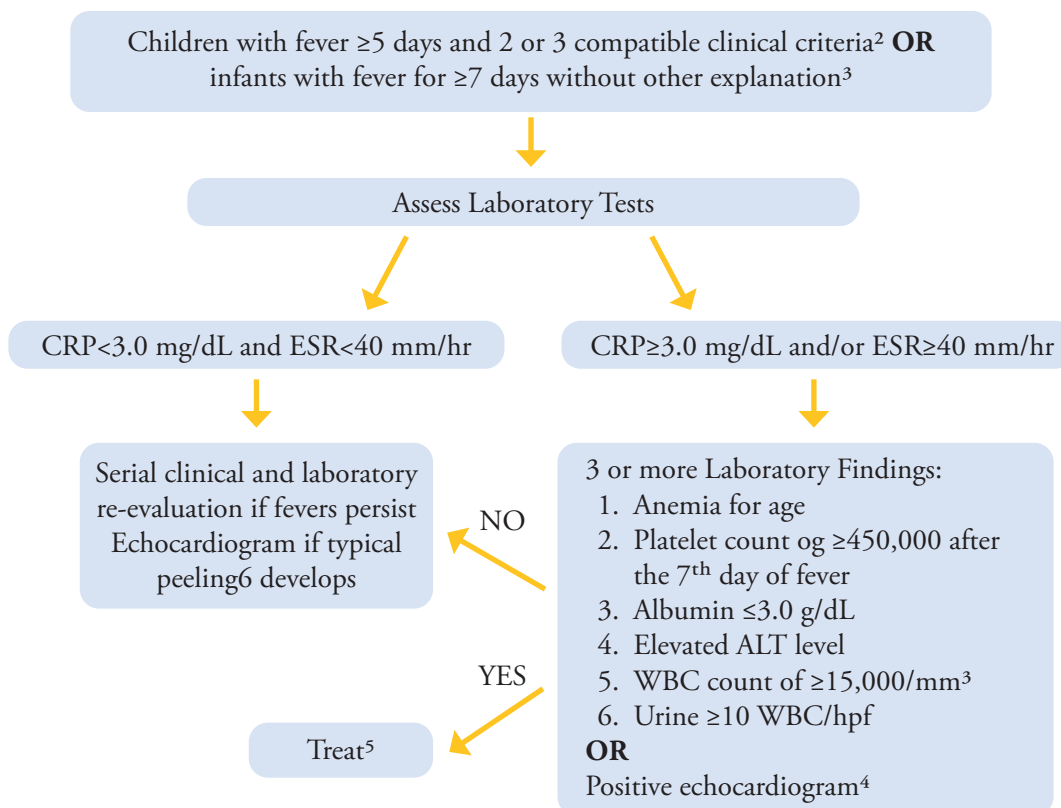
## Treatment

Treatment should begin as soon as possible, ideally within 10 days of when the fever begins. Usually, a child is treated with intravenous doses of gamma globulin (purified antibodies), an ingredient of blood that helps the body fight infection. The child also might be given a high dose of aspirin to reduce the risk of heart problems.

## Evaluation and Testing

If Kawasaki disease is suspected, the doctor may order tests to monitor heart function (such as an echocardiogram) and may take blood and urine samples to rule out other conditions, such as scarlet fever, measles, Rocky Mountain spotted fever, juvenile rheumatoid arthritis or an allergic drug reaction.

## Evaluation of Suspected Incomplete Kawasaki Disease (KD)



\* Infants  $\leq 6$  months old on day 7 of fever without other explanation should undergo laboratory testing and, if evidence of systemic inflammation is found, an echocardiogram, even if the infants have no clinical criteria.

\*\*Supplemental laboratory criteria include:

- Albumin  $\leq 3.0$  g/dL
- Anemia for age
- Elevation of alanine aminotransferase
- Platelets after 7 days of fever  $\geq 450,000/\text{mm}^3$
- White blood cell count  $\geq 15,000/\text{mm}^3$
- Urine  $\geq 10$  white blood cells/high-power field

\*\*\*Echocardiogram is considered positive for purposes of this algorithm if any of 3 conditions are met:

- Z score of LAD or RCA  $\geq 2.5$
- Coronary arteries meet criteria for aneurysms
- $\geq 3$  other suggestive features exist, including perivascular brightness, lack of tapering, decreased LV function, mitral regurgitation, pericardial effusion, or Z scores in LAD or RCA of 2 – 2.5

## Referrals and Consultations

Online: [NationwideChildrens.org/HeartCenter](https://www.nationwidechildrens.org/HeartCenter) or [NationwideChildrens.org/InfectiousDisease](https://www.nationwidechildrens.org/InfectiousDisease)

Phone: (614) 722-6200 or (877) 722-6220 | Fax: (614) 722-4000

Physician Direct Connect Line for 24-hour urgent physician consultations: (614) 355-0221 or (877) 355-0221.

## Follow-up Care

- After being discharged from the hospital, the child generally needs to follow up with their pediatrician within a week.
- Children will be prescribed a low dose of aspirin to be taken for six to eight weeks. Since aspirin can cause gastrointestinal bleeding and other problems, parents should watch for warning signs, such as a stomachache or blood in the stool. Aspirin can cause children to bruise easily, so certain activities should be avoided.
- Parents should also make sure the child is not exposed to anyone with the flu or chicken pox to avoid the risk of Reye's syndrome, which has been linked to aspirin use in these illnesses. Children should also receive the flu vaccine.
- The arthritis from KD is always temporary, but can be uncomfortable. If a child is having difficulty walking or is in pain, the physician may prescribe an anti-inflammatory medication. Physical therapy may also be helpful, or parents may receive a list of exercises to do at home.
- Parents should take their child's temperature, if the child has symptoms of fever, and alert their physician.
- After the child leaves the hospital, blood and urine tests will be ordered to make sure levels return to normal. A cholesterol test is also recommended about a year after the initial diagnosis.
- A low-fat, heart-healthy diet is recommended for all American children. They should also get regular exercise and avoid exposure to secondhand cigarette smoke.

## Guidelines for Risk Stratification for Myocardial Infarction for Long-term Follow-up of Children with Kawasaki Disease

	Risk level	Medical therapy	Physical activity	Follow-up schedule
<b>I</b>	No CA changes at any stage of illness	None after first 6-8 weeks	No restrictions after first 6-8 weeks	CV risk assessment, counseling at 5-year intervals
<b>II</b>	Transient CA ectasia (resolves within 8 weeks)	None after first 6-8 weeks	No restrictions after first 6-8 weeks	CV risk assessment, counseling at 3 to 5-year intervals
<b>III</b>	1 small-medium CA aneurysm in 1 major coronary artery	Low-dose aspirin (3-5 mg/kg per day) until documented regression of aneurysm	Age <11 years: No restrictions after first 6-8 weeks Age 11-20 years: Restriction guided by results of cardiac testing	Annual cardiology follow-up; invasive testing at discretion of cardiologist
<b>IV</b>	≥ 1 large or giant CA aneurysm, or multiple or complex aneurysms in same CA without obstruction	Long-term antiplatelet therapy AND warfarin or low-molecular-weight heparin for those with giant aneurysms	Contact and high-risk sports should be avoided because of the risk of bleeding; other restrictions guided by results of cardiac testing	Cardiology follow-up twice per year; invasive testing at discretion of cardiologist