What is Kawasaki Disease?

Kawasaki disease (KD), also known as Kawasaki syndrome or mucocutaneous lymph node syndrome, is an illness that affects children. It causes inflammation of the blood vessels, particularly the coronary arteries. The average age of those affected is two years — about 77% are younger than five and boys are 1.5 times more likely than girls to get KD. The remaining 25% of cases are older children, adolescents and teens.

KD is seen in all races and ethnicities, but it is more common among children of Asian descent, regardless of where they live.

What are the classic signs of Kawasaki disease?

Kawasaki disease is diagnosed when a patient runs a fever of at least 100.4°F and above for at least five days. The fever is accompanied by at least four of these five symptoms:

- A rash over the torso and extremities.
- Swollen, red hands and feet. Peeling of the skin on the fingers and toes occurs in the second and third weeks of the illness.
- Bloodshot eyes.
- Swollen lymph glands in the neck, usually on one side and measuring more than 1.5 centimeters.
- Redness and swelling of the mouth, lips, throat and tongue. “Strawberry” tongue — the tongue is bumpy and red with enlarged taste buds.

There are cases of “incomplete KD.” In these instances, children have fever and inflammation but do not have all the symptoms.

What causes it?

We don’t know what causes KD. It is not spread from person to person. Rarely more than one child in a family develops it. This may suggest a genetic predisposition for KD.

How does Kawasaki disease affect the heart?

Without prompt treatment, KD can damage the coronary arteries and the heart muscle in as many as 1 in 4 children.

Over the first few weeks, a weakening of a coronary artery can result in enlargement of the vessel wall (aneurysm). A blood clot can form in this weakened area and block the artery. If this happens, the patient can have a heart attack.

When the illness begins, the heart muscle can be inflamed, affecting the heart’s ability to squeeze. Heart rhythm and heart valve problems may occur, but they are rare. While some of this can resolve, the damage to the coronary arteries can be lifelong.
What is Kawasaki Disease?

How is it diagnosed?
KD is a clinical diagnosis, meaning that a caregiver or health care professional identifies the classic signs of KD. A health care professional can also run some blood and urine tests to rule out other illnesses and look for inflammation, but these tests are not specific for KD.

An ultrasound of the heart (echocardiogram) gives a clear picture of the coronary arteries and how the heart is functioning. This may aid in making a diagnosis.

How is Kawasaki disease treated?
Kawasaki disease is treated in the hospital with intravenous immunoglobulin (IVIG) and aspirin. When given early in the illness, IVIG has been shown to reduce the risk of developing coronary artery abnormalities.

If a child is at high risk for coronary artery abnormalities or continues to have a fever after IVIG, there are other medications, such as steroids or infliximab, that can be given.

Aspirin in moderate doses is used in the acute phase to help control fever. After the fever is gone, aspirin is given in lower doses to help prevent blood clots. Aspirin is stopped after 4 to 6 weeks if the child has normal coronary dimensions.

Will my child have long-term issues?
With proper treatment, most children with Kawasaki disease who don’t develop aneurysms recover completely. They don’t typically require long-term follow-up. But it may take one to two months for them to be back to normal. Rest is very important.

Children with KD with aneurysms should have long-term follow-up with a pediatric cardiologist and their care should be transitioned to an adult cardiologist when they become adults.

Most important, people with KD and documented and persistent moderate or large aneurysms require lifelong cardiology follow-up with yearly or biyearly assessments.

HOW CAN I LEARN MORE?

1. Call 1-800-AHA-USA1 (1-800-242-8721), or visit heart.org to learn more about heart disease and stroke.
2. Sign up for our monthly Heart Insight e-news for heart patients and their families, at HeartInsight.org.
3. Connect with others sharing similar journeys with heart disease and stroke by joining our Support Network at heart.org/SupportNetwork.
4. Contact the Kawasaki Disease Foundation at kdfoundation.org.

Do you have questions for your doctor or nurse?
Take a few minutes to write down your questions for the next time you see your health care professional.

For example:

What can I do to help my child deal with Kawasaki disease?

We have many other fact sheets to help you make healthier choices to reduce your risk for heart disease, manage your condition or care for a loved one. Visit heart.org/AnswersByHeart to learn more.