What is Kawasaki Disease?

Kawasaki Disease (KD) is a childhood illness that primarily affects young children. It is characterized by inflammation of the blood vessels throughout the body. About 75 percent of patients are under five years old, but it can affect older children, babies and teenagers, as well. KD is more common in boys than girls, and the majority of cases are diagnosed in the winter and early spring. It is not contagious.

KD gets its name from Tomisaku Kawasaki, a Japanese physician who first described the illness in the medical literature in 1967. In the United States, KD affects people in all racial and ethnic groups, although it is more prevalent among Americans of Asian and Pacific Island descent. About 4,200 children are diagnosed with KD in the U.S. each year.

Although the vast majority of patients who receive timely treatment recover completely, a small percentage develop heart problems. KD can cause abnormal changes in the coronary arteries and other areas of the heart. The most serious health threat is an aneurysm, which is a ballooning of the blood vessel wall. That's why the right diagnosis and timely treatment are so important. KD is the leading cause of acquired heart disease in children.

Early symptoms of KD include:

- 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, which appears 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

Understandably, children with these symptoms are extremely uncomfortable and irritable.

Any parent whose child has persistent fever and any of these symptoms should take the youngster to the doctor immediately.

During the first and second week of illness, other symptoms may appear. A telltale sign of KD is that skin on the fingertips and toes starts to peel. Children may also develop temporary arthritis that causes pain in joints throughout the body, an enlarged gallbladder, temporary hearing loss, abdominal pain, vomiting and diarrhea.

What causes KD?

In some ways, KD is a mysterious illness. The precise cause is unknown. Researchers believe that some kind of infection, as yet undefined, or an unknown environmental factor may trigger the illness in individuals who have a genetic predisposition. Various reports have linked KD to a number of possible causes, including a prior respiratory illness, exposure to carpet-cleaning chemicals, use of a humidifier or living near a stagnant body of water. However, there is no firm evidence that any of these factors causes KD.

What is the prognosis of children who’ve had KD?

Most children will make a complete recovery and resume normal activities within four to eight weeks. Children in whom heart problems have been detected will need to be closely monitored by a cardiologist.

The long-term prognosis depends on the degree of coronary artery damage early in the illness. The outlook for children who have not developed cardiac abnormalities is very good. Follow-up studies of children who never had aneurysms do not show an increased incidence of early heart disease later in life.

Researchers around the country are conducting studies to better understand the causes, best treatments and long-term effects of KD.

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The Kawasaki Disease Foundation (KDF) is a national nonprofit organization dedicated to advancing Kawasaki Disease issues by raising awareness, supporting families and promoting research. It represents a true collaboration of parents and medical professionals.

The Foundation strives to increase early detection and treatment of Kawasaki Disease, and provides information and support for families through a nationwide telephone and e-mail help network.

Support volunteers are generally parents of children who have had KD.

To contact a support volunteer, please visit our web site: www.KDFoundation.org or send an email to us at info@KDFoundation.org

Ask us about Kawasaki Disease.
How is KD diagnosed?
As of right now, there is no specific laboratory test, such as a blood test or throat culture, to diagnose KD. A doctor makes the diagnosis based on the child's symptoms. Diagnostic criteria include high fever for four or five days, along with four of the symptoms mentioned previously. Doctors also order blood work. Signs of inflammation in the blood are another clue that a child may have KD.

Although most pediatricians recognize KD in children who have the classic symptoms, it is more difficult to make the diagnosis in patients who do not fit the typical profile. The problem is that the signs and symptoms of KD occur in other illnesses. The disease is most frequently misdiagnosed in babies younger than one year old and in teenagers.

The American Heart Association has issued guidelines for physicians in diagnosing KD based on the recommendations of a committee of highly regarded experts. The proper diagnosis is critical. Treatment within 10 days after fever appears is essential to decrease the risk of heart problems.

How is KD treated?
Children are hospitalized and receive an intravenous treatment called gamma globulin, a biological product that comes from donor blood and contains antibodies. The treatment, also called immunoglobulin or IVIG, is given for eight to 12 hours. A child’s temperature usually returns to normal after treatment. If fever persists, the child may receive a second course of IVIG treatment.

Although we don’t know exactly how it works, gamma globulin decreases the risk of heart problems that pose the greatest threat to patients with KD. With treatment, only one to five percent of patients develop coronary abnormalities. However, 15-25 percent of patients who are not treated within 10 days of illness onset develop aneurysms. Children are also given high doses of aspirin during the first week or two, and it is very important to call the doctor if the youngster develops a fever. Children may require a second intravenous treatment if they have fever. If parents have any questions, they should not hesitate to call their doctor or the hospital.

Once children recover, can they get KD again?
A recurrence is uncommon. Only one to three percent of children come down with KD a second time.

What can parents expect after a child comes home from the hospital?
Upon discharge, the hospital usually gives parents a list of instructions. They generally follow up with their pediatrician within a week after coming home from the hospital.

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 stomach ache or blood in the stool. Aspirin can also 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.

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.

To one or two weeks after the illness starts, children may have peeling on their fingers and toes. Although it is usually not painful, the new skin underneath can be tender. Overall, a child’s skin may be dry for several weeks and you can use an unscented lotion to moisturize the skin.

Parents should take their child’s temperature daily for the first week or two, and it is very important to call the doctor if the youngster develops a fever. Children may require a second intravenous treatment if they have fever. If parents have any questions, they should not hesitate to call their doctor or the hospital.

Is the heart checked after the child leaves the hospital?
It is recommended that echocardiograms be performed to detect aneurysms and other abnormalities at the time of diagnosis, two weeks after illness onset, and then six to eight weeks after the illness started. Children who’ve had changes in their coronary arteries may have more frequent tests. Even when the tests are normal, many doctors also prescribe a follow-up echocardiogram, which is given one year after the child came down with KD. Some physicians also recommend another echocardiogram five years after the initial diagnosis.

Children whose tests show heart problems must be monitored much more closely.

Are any other follow-up tests recommended?
After the child leaves the hospital, blood and urine tests will be ordered to make sure they return to normal. A cholesterol test is also recommended about a year after the initial diagnosis.

Should children who had KD have a heart-healthy diet, even if they have not had coronary artery damage?
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.

Needless to say, they should not start smoking when they get older.