

The following are some of the questions most frequently asked by parents of children who suffer from Kawasaki Disease. For a copy of this handout and other information on Kawasaki Disease, visit our website and the website for the KD Foundation, a national parent organization committed to increasing awareness and education about KD:

<http://www.pediatrics.ucsd.edu/kawasaki>

<http://www.kdfoundation.org>

WHAT IS KAWASAKI DISEASE?

Kawasaki Disease is an unusual illness characterized by inflammation of blood vessels throughout the body. It is accompanied by the following symptoms: fever; rash; swelling of the hands and feet; bloodshot eyes; irritation and redness of the mucous membranes of the mouth, lips, and throat; and swollen lymph nodes in the neck. The immediate effects of Kawasaki Disease may not be serious but, in some cases, long-term complications including damage to the coronary arteries (vessels that supply blood to the heart muscle) and heart muscle may result.

Kawasaki Disease affects children almost exclusively; most patients are under 5 years of age. For reasons still unknown, males acquire the illness almost twice as often as females.

The disease is named after the Japanese pediatrician, Tomisaku Kawasaki, who described this particular pattern of signs and symptoms in 1967. Since then, Kawasaki Disease has been found to occur most frequently among Japanese children. In the United States, the disease has been reported in all racial and ethnic groups but occurs most often among children of Asian-American descent. Kawasaki Disease is not a rare illness and the exact number of cases that occur in the United States has not been determined. We estimate that there are between 4,000-5,000 diagnosed cases of Kawasaki disease each year in the U.S. It is estimated that the disease attacks 15-20 out of every 100,000 children less than 5 years of age. The disease can occur in clusters or localized outbreaks - usually in the winter and spring.

WHAT CAUSES KAWASAKI DISEASE?

To date, no cause of Kawasaki Disease has been identified. Most experts agree that an infectious cause (such as a virus or bacteria) is likely, although a hereditary tendency also exists and explains why the disease occurs more frequently among children of Asian ancestry. Younger brothers and sisters of a KD patient have a 10-fold increased risk of KD because of a shared genetic predisposition. When children with Kawasaki Disease grow up, their children are at higher risk of developing KD. At the present time there is no evidence that the disease is contagious.

WHAT ARE THE SIGNS AND SYMPTOMS?

Fever and irritability often occur first. The fever has a rapid onset and fluctuates from moderate (101° to 102° F) to high (above 103°). Lymph nodes in the neck may become swollen. A rash usually appears early in the illness; some patients may develop an accentuated rash in the groin. Often vivid red in appearance, the rash is composed of either poorly defined spots of various sizes or larger masses of merging spots. Fever continues to rise and fall, possibly for up to three

weeks. Bloodshot eyes (conjunctival injection), usually without discharge, develop during the first week of illness.

A child's tongue may be red and show small, raised bumps (papillae), called "strawberry tongue" because the enlarged papillae resemble the seeds on a strawberry's surface. The lips become dry and cracked and often take on a bright red color. Mucous membranes of the mouth turn a darker red than usual.

The palms of the hands and soles of the feet often turn bright red. Hands and feet can swell. Occasionally, a child may develop a stiff neck. The child usually has great difficulty getting comfortable and may be very irritable. Joint symptoms include stiffness and pain in the hips and knees and sometimes the small joints in the hands. The child may refuse to walk. Many parents also notice hoarseness during the acute illness.

When the fever subsides, the rash, red eyes and the swollen lymph nodes usually disappear. Skin starts to peel around the toenails and fingernails, often beginning during the third week of illness. The skin on a hand or foot may peel off in large pieces or even a single piece (much as a snake sheds its skin). Knees, hips, and ankles can become more inflamed and painful.

Occasionally, joint pain and inflammation persist after other symptoms have disappeared. Transverse (horizontal) depressed lines on fingernails and toenails, which occur during the illness, may be visible for months afterward until the nails grow out. Hair loss may occur. Eczema may flare and require treatment. Psoriasis may appear for the first time.

HOW DOES A DOCTOR DETERMINE IF A CHILD HAS KAWASAKI DISEASE?

A physician makes a diagnosis of Kawasaki Disease after carefully examining a child, observing signs and symptoms, and after ruling out the possibility of other diseases that can cause similar signs. Blood tests are used to detect mild anemia, a white-blood-cell count above normal, and an elevated erythrocyte sedimentation rate which indicates blood vessel inflammation. A sharp rise in the number of platelets, the major clotting element in the blood, may also be detected. Urine tests may reveal unusual white blood cells in the urine. Irregular heart rhythms (arrhythmias) and evidence of heart muscle strain, indicating involvement of the heart, can be detected by an electrocardiogram (EKG). Echocardiography (a sound wave diagnostic test of heart and blood vessel structure and function) is necessary to evaluate possible damage to the heart or large blood vessels.

HOW IS KAWASAKI DISEASE TREATED?

A high dose of intravenous gamma globulin (IVIG, a protein fraction of human blood) is the treatment of choice for patients with Kawasaki Disease. This treatment is most effective in reducing inflammation and preventing coronary artery damage if it is started within the first 10 days of illness. High doses of aspirin are also given with gamma globulin during the acute phase of the illness until the fever subsides. Complications from treatment are rare. Viruses such as HIV (the AIDS virus) and Hepatitis C virus cannot be transmitted by currently available IVIG products. Occasionally, chills, fever, and a drop in blood pressure may occur during the infusion. This is treated by interrupting the infusion and giving an antihistamine before restarting. High

doses of aspirin may sometimes cause abdominal pain, gastrointestinal bleeding, and ringing in the ears. Aspirin should be discontinued if any of these signs or symptoms appear. Reye Syndrome is a rare complication of aspirin therapy that can occur in children exposed to chicken pox or influenza virus while they are taking high doses of aspirin. Low dose aspirin carries no risk of Reye Syndrome.

If diagnostic tests reveal the presence of an aneurysm (dilated segment of the coronary artery) or any other heart or blood vessel abnormality, medical or surgical treatment may be needed. Your doctor may recommend that a cardiologist (a physician who specializes in heart problems) monitor a heart or blood vessel problem for several years following recovery from Kawasaki Disease.

WHAT ARE THE CONSEQUENCES IF THE DISEASE IS NOT PROPERLY TREATED?

Fever, swollen lymph nodes (also called "swollen glands") in the neck, rash, and mucous membrane inflammation can be extremely uncomfortable and last for 1 to 3 weeks without treatment. With treatment, the fever and other symptoms usually subside within 24 hours.

About 25 percent of children who are affected by the disease develop heart problems in the latter stages of the illness. Damage to large blood vessels which supply the heart muscle, as well as damage to the heart itself, can sometimes occur. A weakening of the large vessels in the heart (coronary arteries) can result in an enlargement or ballooning (aneurysm) of the blood vessel wall.

Full recovery can be expected in most cases, but the possibilities of blood vessel and heart disease in later life remain subjects of medical investigation. Infants less than 1 year old usually become most seriously ill and are at greatest risk for coronary artery damage. Less than 1% of the American children who contract the disease die during the initial illness.

ARE THERE ANY COMPLICATIONS ASSOCIATED WITH KAWASAKI DISEASE?

Heart and blood vessel problems can make Kawasaki Disease unpredictable. Most often these problems are not serious and disappear in time. However, aneurysms of coronary or other large arteries can be serious and may require medical or surgical treatment. Severe blood vessel and heart complications can prove fatal. Late after Kawasaki disease there may be scarring of the blood vessels and heart muscle that can cause new problems in some young adults.

Heart muscle inflammation (cardiomyopathy) and congestive heart failure may accompany the fever. An abnormal and painful accumulation of fluid in the **gall bladder** (hydrops of the gall bladder) resulting in severe abdominal pain sometimes occurs during the period of fever. Inflammation of the membranes around the brain may cause a sterile meningitis. On rare occasions, damage to the nerve involved in hearing can rarely occur and result in **deafness**. Therefore, children with Kawasaki Disease should have a hearing test performed after recovery if there is any question about their hearing. Both **eczema** and **psoriasis** are linked to KD and may present as a skin rash in the weeks following the onset of fever. A temporary **arthritis** of the hips, knees, or ankles is common following resolution of the fever.

WHAT CAN I EXPECT ONCE MY CHILD COMES HOME FROM THE HOSPITAL?

After coming home from the hospital, you may notice that your child continues to be tired and has a poor appetite for about 1 to 2 months. However, unless you have been specifically told otherwise by your doctor, you should not attempt to limit your child's activity or diet.

You should call your doctor immediately, however, if any of the following symptoms occur:

1. Signs of aspirin toxicity (while on high dose aspirin).

This is characterized by the following symptoms:

- a. shallow rapid breathing
- b. complaints of stomach pain
(with or without vomiting blood, which looks like coffee grounds)

2. Return of fever and other signs of Kawasaki Disease (e.g. rash, red eyes - see Signs and Symptoms section above).

Note: Tenderness or swelling of the large joints (elbows, knees) and peeling of the fingertips and toes are a normal part of the recovery, but should resolve after approximately 3 weeks.

Note: Routine live virus immunizations for measles, mumps, and rubella (MMR) and chickenpox (varicella) should be delayed for 12 months following gamma globulin treatment.

CAN MY CHILD CONTRACT THIS DISEASE AGAIN IN THE FUTURE?

Rarely, Kawasaki Disease can reoccur months to years after the initial disease (in Japan, recurrences have been reported in up to 10% of cases). Should the signs and symptoms described earlier in this pamphlet reoccur, call your doctor immediately.

CAN KAWASAKI DISEASE BE PREVENTED?

Unfortunately, at this time, Kawasaki Disease cannot be prevented. However, programs such as the Kawasaki Disease Research Program at UCSD/Rady Children's Hospital San Diego are working in conjunction with researchers throughout the United States and other countries to further understand this mysterious disease.

TO LEARN HOW YOU CAN PARTICIPATE IN KAWASAKI DISEASE RESEARCH, CONTACT THE KAWASAKI DISEASE RESEARCH PROGRAM AT

kdgenetics@ucsd.edu