

What is Kawasaki Disease?

Kawasaki Disease is a disease that causes general inflammation of blood vessels. We do not know the cause of Kawasaki Disease, but we know that it seems to cause the body to generate a strong reaction against itself. Some people feel that a previous infection with Staph or Strep infection may play a role. More specifically, it causes the body to attack its own blood vessels and cause them to weaken and sometimes rupture. Our biggest fear with Kawasaki Disease is that it can sometimes affect the coronary arteries (the blood vessels that feed the muscle of the heart) and some patients can have a heart attack because of a clot and sometimes die because of it.

A Japanese physician first described Kawasaki disease in the mid 1960's. It was first described in the American literature in the mid 1970's. It is much more common in the Japanese population, by a factor of about ten times. The incidence of the disease in America is about 30 per 100,000 children. It is 1.5 times more common in boys than girls. The highest risk group for aneurysm formation of the coronary arteries and death is males under six months of age. There is no Laboratory test to diagnose Kawasaki disease.

We make the diagnosis based on certain clinical findings. These include 1- high fever for several days unresponsive to antibiotics, 2- a characteristic rash, 3- red eyes and mouth, 4- swollen hands and feet, 5- lymph nodes, later they develop peeling of the fingers and toes. Blood work reveals a high white cell count and a high sedimentation rate (an indication of inflammation). The platelet count usually begins to rise after the first week and can reach levels of over one million (normal is less than 300,000) Damage to the coronary arteries is usually not detected until ten days to two weeks after the beginning of the fever and for this reason we usually obtain a base line echo study at the beginning of the diagnosis, a second one at about two weeks out and a third one at 6 to 8 weeks from the beginning of the fever. If no damage is seen after six to seven weeks from the beginning of the fever then it is pretty safe to assume that no damage will occur.

Treatment of Kawasaki disease includes the use of I.V. gamma globulin which is given over one or two days and high dose aspirin to reduce the inflammation. Once the fever is gone we then continue with low dose aspirin for six weeks to prevent clotting of blood within the coronary arteries. The incidence of damage to coronary arteries (aneurysm formation) when gamma globulin is used is less than 5%. With out gamma globulin the incidence is about 20%. The reason I.V. gamma globulin works is that it shuts down the production of the body's own antibodies against its own blood vessels which is felt to be the cause of the findings of Kawasaki disease. Fortunately, even among patients who develop aneurysms over half will get better. For those who continue to show signs of damage to the coronary arteries aspirin therapy is suggested for life. Very rarely is it necessary to do surgery to open the coronary arteries. This is very risky in the small babies. The type of therapy, follow-up or restrictions for exercise will depend on the damage to the coronary arteries. It is probably a good idea to obtain an exercise stress test when the child is 10 to 12 years old. These patients are probably at higher risk of coronary artery disease as adults.