KAWASAKI'S DISEASE

Immune-Mediated Vasculitis in Children

Kawasaki's disease, which primarily affects infants and small children, is characterized by a sudden onset of fever, rash, and vasculitis.

Kawasaki disease is an acute self-limited immune-mediated form of vasculitis. First described in Japan in 1967, Kawasaki’s disease is known to occur in both endemic and community-wide epidemic forms in North America, South America, Europe, and Asia, affecting children of all races.

Prevalence

Kawasaki’s disease is most prevalent in Japan with an annual incidence of about 112 cases per 100,00 children less than 5 years old. In the United States more than 4,000 children are hospitalized with Kawasaki’s disease annually, with a median age of 2 years and a predominance in Americans of Asian and Pacific Island descent. The incidence of Kawasaki’s disease in the United States is intermediate in Hispanics and African Americans and lowest in white children. When Kawasaki’s disease appears in older children, cardiac complications are more likely to occur and may be related to a late diagnosis.

Children who have siblings, especially twins, with Kawasaki’s disease have a higher disease risk than other children. Because children with a parent who had Kawasaki’s disease as a child also have a higher disease risk, genetic factors are suspected of contributing to disease development.

Symptoms and Signs

Children with Kawasaki’s disease may have a variety of different symptoms including an abrupt onset of fever, nausea, vomiting, gallbladder disease, conjunctivitis affecting both eyes, reddened lips and oral mucosa, rash, enlarged cervical lymph nodes, and changes in the extremities including arthritis. Untreated, up to 25 percent of affected children may develop coronary artery aneurysms, heart attacks, ischemic heart disease and coronary thrombosis (blood clots), making Kawasaki’s disease the most common cause of heart disease in children.

Fever in Kawasaki’s disease typically lasts 5 days or longer and is high spiking and remittent, with peak temperatures generally above 102 degrees F and sometimes above 104 degrees F. With appropriate treatment, fever usually resolves within 2 days. Without treatment, the fever persists for a mean of 11 days although it may last as long as 3-4 weeks or longer.
The rash in Kawasaki’s disease can take different forms, most commonly a nonspecific, diffuse, eruption of flattened papules. The rash usually covers the entire body, including the trunk and extremities, and itching may occur. A reddened “strawberry” tongue may also occur and be confused with strep throat. Cardiovascular abnormalities are the leading cause of long-term morbidity and mortality.

**Laboratory Changes**

Laboratory changes in Kawasaki’s disease include positive tests for perinuclear anti-neutrophilic cytoplasmic antibodies (P-ANCA), increased white blood cell count, anemia, elevated erythrocyte sedimentation rate (sed rate), elevated C-reactive protein (CRP) level, and an increased platelet count.

**Causes and Diagnosis**

The origins of Kawasaki’s disease remain perplexing. Although a specific infectious agent has not been identified in Kawasaki’s disease, an infectious trigger is suspected. Another perplexing fact is that, unlike other forms of vasculitis, patients with Kawasaki’s disease are not easily treated with corticosteroids.

There are no specific diagnostic tests available to diagnose Kawasaki’s disease. Rather, this disease is suspected when the characteristic signs and symptoms are present in small children. An incomplete form of Kawasaki’s disease, causing only a few of the characteristic signs, may occur and be difficult to distinguish from viral as well as Staphylococcus and Streptococcus infections. In the past Kawasaki’s disease may have been misdiagnosed as polyarteritis nodosa with reports of old cases appearing identical to fatal cases of Kawasaki’s disease today.

In addition, symptoms may show up at different times, sometimes not occurring until the initial abrupt onset of symptoms appears to have resolved. Varying degrees of myocardial scarring, thickening and fibrosis may show up later in children with Kawasaki’s disease who had normal echocardiograms early in the course of the illness. In autopsies of children with fatal Kawasaki’s disease, coronary and lymph node changes, including lymph node necrosis, are typical.

**Treatment**

Treatment consists of aspirin therapy and intravenous immunoglobulin therapy. Up to 80 percent of children treated with intravenous immunoglobulins show a good response to therapy. Of the children who do not respond to this therapy, the addition of corticosteroids does not cause improvement.

**Resources:**