

without treatment, approximately 20% to 25% of children develop coronary artery dilation or aneurysm formation. Infants younger than 1 year old are at the greatest risk for heart involvement, although an increased incidence has also been reported in older children, perhaps because of later diagnosis in many.

The etiology of Kawasaki disease is unknown. The illness is not spread by person-to-person contact; however, several factors support an infectious etiologic trigger, possible in a genetically susceptible host. It is often seen in geographic and seasonal outbreaks, with an increased incidence reported in the late winter and early spring ([Newburger, de Ferranti, Fulton, et al, 2015](#); [Newburger, Takahashi, Gerber, et al, 2004](#)).

Pathophysiology

The principal area of concern in Kawasaki disease is the cardiovascular system. During the initial stage of the illness, extensive inflammation of the arterioles, venules, and capillaries is evident, resulting in many of the clinical symptoms. In addition, segmental damage to the medium-sized muscular arteries, mainly the coronary arteries, can occur, resulting in the formation of coronary artery aneurysms in some children. Death is very rare in Kawasaki disease (<0.17% of cases) and is usually the result of myocardial ischemia from coronary thrombosis during the first few months of illness or years later from severe scar formation and stenosis in coronary aneurysms ([Wilder, Palinkas, Kao, et al, 2007](#)).

Clinical Manifestations

Because no specific diagnostic test exists for Kawasaki disease, the diagnosis is established on the basis of clinical findings and associated laboratory results ([Box 23-12](#)). These criteria should be used as guidelines. It is important to note that many children with Kawasaki disease do not fulfill standard diagnostic criteria, and infants in particular often have an incomplete presentation. It is therefore important to consider Kawasaki disease as a possible diagnosis in any infant or child with prolonged fever that is unresponsive to antibiotics and is not attributable to another cause.

Box 23-12