ARTICLE

Preventing Long-Term Cardiac Damage in Pediatric Patients With Kawasaki Disease

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ABSTRACT

Kawasaki disease is currently the leading cause of long-term cardiac damage in pediatric patients in the United States. Kawasaki disease is diagnosed based on symptomatology and by ruling out other etiology. There is a significant need for an improved, standardized treatment protocol for patients diagnosed with Kawasaki disease and a more rapid initiation of treatment for these patients. Decreasing the cardiac damage caused by Kawasaki disease with timely diagnosis and treatment needs be a principal goal. J Pediatr Health Care. (2016) ■, ■-■.

KEY WORDS

Cardiac damage, coronary artery aneurysm, fever, Kawasaki disease, pediatrics

Treatment recommendations for patients with Kawasaki disease include the use of high-dose aspirin and intravenous immunoglobulin (IVIG) infusion. In patients who are resistant to IVIG, studies are reporting promise with the use of infliximab. There is also research that shows promise with the use of abciximab in patients who develop coronary artery aneurysm; studies are showing resolution of these coronary artery aneurysms with the use of abciximab. Treatment needs to be initiated within the first 10 days of fever to be most effective and for the best chance of reducing the risk of long-term cardiac damage.

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Conflicts of interest: None to report.

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BACKGROUND

Kawasaki disease is the number one cause of acquired cardiac damage in children in the United States. Studies have shown a 56% increase in endothelial dysfunction in patients previously affected by Kawasaki disease compared with those with no prior history of disease. Kawasaki disease is defined as "an acute febrile illness of unknown etiology that primarily affects children younger than 5 years of age" (Centers for Disease Control and Prevention, 2013). Kawasaki disease is a form of vasculitis that causes inflammation of blood vessels throughout the body and can affect the arteries, veins, capillaries, and coronary arteries (National Heart, Blood, and Lung Institute, 2011). Kawasaki disease was first identified in Japan by Tomisaku Kawasaki in 1967 (CDC, 2013). It affects children of Asian descent at an incidence of approximately 10 in 1,000 but can affect children of any ethnic background. Boys are twice as likely as girls to be diagnosed with Kawasaki disease (Seattle Children's, 2015). Although more cases are reported annually in Asian countries, especially Japan, there are approximately 7,000 cases of Kawasaki disease reported annually in the United States (Seattle Children's, 2015). Scheinfeld

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(2015) reports that the annual incidence in the United States per 100,000 children under the age of 5 years is approximately 32.5 cases for "Americans of Asian and Pacific Island descent, 16.9 cases for non-Hispanic African Americans, 11.1 cases for Hispanics, and 9.1 cases for whites" (p. 5).

The primary clinical signs associated with Kawasaki disease include persistent fever greater than 102°F; nonexudative bilateral conjunctivitis; anterior uveitis; erythema, fissures, and crusting of the lips; diffuse mucosal injection of the oropharynx; strawberry tongue; erythema and edema on the palms of hands and soles of feet; polymorphous nonvesicular rash that may be concentrated on the trunk, groin, or lower extremities; desquamation in the genital region, hands, and feet, more pronounced around nails, palms, and soles; perianal erythema; lymphadenopathy of a unilateral, nonsuppurative cervical node measuring approximately 1.5 cm; and pain and swelling in joints, often bilateral.

The risk associated with Kawasaki disease is that vasculitis can lead to myocardial infarction, thrombus formation, aneurysms, and/or coronary artery dilation (Hockenberry & Wilson, 2010). It is estimated that coronary artery aneurysms occur in 15% to 25% of children with Kawasaki disease who are untreated, which can lead to myocardial infarction, ischemic heart disease, and sudden cardiac death (Seaton & Kharbanda, 2015). These cardiac effects can be seen during the acute stage of the illness or years to decades after resolution of Kawasaki disease (Hockenberry & Wilson, 2010). However, there are no accurate data regarding the percentage of patients who suffer from cardiac disease later in life that is related to having had Kawasaki disease in childhood. Kawasaki disease is now the leading cause of acquired cardiac disease in children in the United States, surpassing rheumatic heart disease (Seaton & Kharbanda, 2015). Cardiac death associated with Kawasaki disease is almost results from cardiac sequelae (Seaton & Kharbanda, 2015). Although rarely fatal, the peak mortality rate occurs within 15 to 45 days after the onset of fever (Seaton & Kharbanda, 2015). To reduce the risk of long-term cardiac damage associated with Kawasaki disease, it is essential that providers are able to effectively and appropriately assess the clinical presentation and laboratory results associated with Kawasaki disease, rule out other pathologies, and initiate prompt and appropriate treatment.

LITERATURE REVIEW

Diagnosis

With no diagnostic test available for Kawasaki disease, diagnosis is made by physical examination and history and through a process of ruling out other pathologies. The current guidelines for the diagnosis of Kawasaki disease in the United States include the presence of fever for five or more days along with four of the five following criteria: bilateral bulbar, nonsuppurative conjunctivitis; cervical lymphadenopathy, often greater than 1.5 cm; polymorphous, nonvesicular, noncrusting rash; erythema of the lips, lip fissures, "strawberry" tongue, or diffuse erythema of the oropharynx; erythema and edema of the palms of the hands and soles of the feet; and peeling of the skin on the fingertips (Eleftheriou et al., 2014). Although not diagnostic, elevated C-reactive protein (CRP) level, erythrocyte sedimentation rate, and/or leukocytosis may also be seen in patients with Kawasaki disease (Eleftheriou et al., 2014).

It is possible to diagnose incomplete Kawasaki disease, in which patients do not fulfill the criteria for complete Kawasaki disease but may still be at risk for coronary artery aneurysm (Eleftheriou et al., 2014). Incomplete Kawasaki disease is diagnosed when patients present with some of the features of Kawasaki disease and an elevated CRP level, erythrocyte sedimentation rate, or leukocytosis (Eleftheriou et al., 2014). Patients in this group should undergo echocardiography, which may or may not indicate coronary vasculitis. Although the presence of coronary vasculitis confirms the diagnosis of Kawasaki disease, a negative echocardiography result does not exclude the diagnosis of Kawasaki disease (Eleftheriou et al., 2014).

Eleftheriou et al. (2014) state that a diagnosis of Kawasaki disease should be made in the presence of five of the six diagnostic criteria when present before Day 5 of fever, when coronary artery aneurysm or coronary artery dilation is present, or there is evidence of continued elevation of inflammatory markers when no other diagnosis has been made and there is a high suspicion of Kawasaki disease. One presenting symptom that is almost often present, but is not one of the diagnostic criteria, is irritability. Irritability in Kawasaki disease is significantly increased and is markedly increased from the child's normal temperament when ill and should be considered an important sign when evaluating for Kawasaki disease (Eleftheriou et al., 2014).

Coronary Abnormalities

A literature review on the subject of preventing long-term cardiac damage in pediatric patients diagnosed with Kawasaki disease through early diagnosis and treatment showed supporting evidence and recommendations for various treatment modalities. Rowley (2012) again confirms that Kawasaki disease remains the primary cause of acquired cardiac disease in pediatric patients in developing nations. With an unknown causative agent, Kawasaki disease continues to cause diagnostic difficulties for providers who may see only one or two patients with the disease per year in practice (Rowley, 2012).

Rowley (2012) supports the need for diagnosis and treatment of Kawasaki disease before the 10th day of