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Kawasaki Disease in Pediatric Patients

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Background

Kawasaki Disease (KD) is an idiopathic, multisystem disorder. It is characterized by vasculitis of the arteries, capillaries, and veins and typically affects children 5 years old or younger (Lubin, Weinreb, Lobry, & Duke, 2015). This inflammation of the blood vessels makes Kawasaki disease the leading cause of acquired heart disease in children, with 20% of those affected developing coronary artery aneurysms, cardiac arrhythmias, and heart failure (Lubin et al., 2015). KD occurs in all races, but is predominately seen in children of Japanese descent. Diagnosis is based on criteria including presence of fever for 5 days, bilateral conjunctivitis, erythema of the lips and oral mucosa, changes in extremities, rash, and cerebral lymphadenopathy (Sehgal, Chen, & Ang, 2015). Treatment of KD typically involves high-dose aspirin and intravenous immunoglobulin (IVIG) while hospitalized and low-dose aspirin until inflammatory markers decrease (Sehgal et al., 2015).

Signs & Symptoms

There is no specific diagnostic test available for KD and diagnosis is usually made using the clinical criteria adopted by the American Heart Association. At least 5 of the following symptoms must be seen:
- Fever persisting greater than 5 days
- Changes in extremities, including indurative angioneurodesia and desquamation
- Polyynymphotic exanthema (diffuse red rash)
- Bilateral bulbar conjunctival injection without exudate
- Changes to the lips and oral cavity, including pharyngeal injection, dry fissured lips, and/or strawberry tongue
- Acute nonpurulent cervical lymphadenopathy
- Elevated inflammatory markers (C-reactive protein and erythrocyte sedimentation rate) (Jamieson & Singh-Grewal, 2013)

Underlying Pathophysiology

- A specific agent has not been identified, however KD is thought to be triggered by an infectious agent that causes an immune response in a genetically susceptible host.
- An infectious etiology is also supported by the fact that KD is seen predominantly in the winter and spring, found mostly in children, and is self-limiting similar to other viral diseases.
- The vasculitis of KD reveals an infiltration of macrophages, neutrophils, and CD11b cells (Dimitriadis, Brown, & Gedalia, 2014).
- Pro-inflammatory cytokines that are released are directly related to the fever, mucosal involvement, and desquamation seen in KD (Dimitriadis, Brown, & Gedalia, 2014).
- These cytokines then target the endothelial cells and result in a cascade of events that cause vascular damage. In severely affected vessels, the media develops inflammation with necrosis of smooth muscle cells (Medscape, 2015).
- Over the next few weeks to months, the active inflammatory cells are replaced by fibroblasts and monocytes, and fibrous connective tissue begins to form within the vascular wall.
- The vessel wall eventually becomes narrowed or occluded owing to stenosis or a thrombus. Cardiovascular death may occur from myocardial infarction secondary to thrombosis of a coronary aneurysm or from rupture of a large coronary aneurysm (Medscape, 2015).

Significance of Pathophysiology

- The period of the greatest vascular damage is due to the increase in the serum platelet count, and this is the point of the illness when the risk of death is most significant (Medscape, 2015).
- Also, pathologic findings should change the way practitioners think about “regression” of coronary artery aneurysms after KD. The lumen of saccular aneurysms seen in some patients with KD becomes smaller over time because of deposition of sequential layers of thrombus; this decrease in lumen size does not necessarily represent “healing” of the arterial wall (Rowley, 2012).
- Decrease in size of the lumen of fusiform aneurysms may result from thrombus formation or from IAMP stenosis. Although it is unlikely that aneurysmal arteries return to normal, nonneuronal arteries that develop mild dilatation as a result of edema and minimal inflammatory infiltrate likely can return to normal or near normal (Rowley, 2012).
- Understanding the underlying pathophysiology leads to a better understanding of the potential dangers facing KD patients with severe coronary artery abnormalities over time (Rowley, 2012).
- Studies show that risk factors for the development of coronary artery abnormalities in patients with KD and prevalence of coronary disease were lower in children who were treated before the fifth illness day compared with after the fifth illness day was reported (Rowley, 2012).

Epidemiology/Etiology

KD has a higher incidence in Japanese-Americans, as well as an increased risk in the siblings of those affected. This suggests that host genetic factors are important in determining the abnormal immunological response to the infectious triggers. KD also predominantly affects young children; 80% of cases occur between the ages of 6 months and 4 years and has a male predominance (Kim, Curtis, Cheung, & Burgen, 2013). KD also has a peak incidence in the winter and spring. Over time, a rising incidence has been observed worldwide, possibly due to heightened awareness and recognition of the disease (Jamieson & Singh-Grewal, 2013).

Implications for Nursing Care

Treatment for KD includes administration of intravenous immunoglobulin and aspirin.
- Intravenous immunoglobulin: The mechanism of action of IVIG remains unknown, however a single dose of 2g/kg given over 10–12 hours, paired together with aspirin within 10 days of fever onset results in rapid resolution of clinical symptoms in 80–90% of patients and has been shown to reduce the risk of coronary disease from 20–25% to about 2–4% (Patel & Shulman, 2015).
- Aspirin: Anti-inflammatory doses of 80–100 mg/kg/day is used in the acute phase, followed by lower antplatelet doses of 3–5 mg/kg/day after defervescence (Patel & Shulman, 2015).
- Other therapies: In patients who do not respond to primary therapy with IVIG and aspirin, the best management approach remains uncertain. Additional therapy options include a second dose of IVIG, pulse prednisone therapy, and infliximab. Most recently, the use of calcineurin inhibitors, particularly cyclosporine, has been used for treatment of patients who failed multiple other therapies. Because the clinical symptoms of KD are self-limiting, it is difficult to determine whether case reports of patients who apparently responded to rescue therapies represent true clinical responses (Rowley, 2012).

Conclusion

Kawasaki Disease is the most common cause of acquired heart disease for children in the United States. Primary health providers play an important role in the early detection and diagnosis of children with KD. Timely diagnosis and administration of treatments has been shown to improve outcomes and reduce the risk of cardiac complications.

References