Kawasaki Disease

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Background

Definition

Kawasaki disease (KD) is a rare, self-limiting generalized vasculitis that primarily affects the coronary arteries. While the exact etiology is unknown, it is considered to be the most common cause of coronary artery disease in young children.

Etiology

1. While the etiology of KD has yet to be fully elucidated, the leading theories suggest two main categories as potential triggers: environment toxins and infections [1–20]. Interestingly, the common pathway for these KD triggers appears to be the respiratory tract. Given the seasonal patterns of KD in Japan, Hawaii, and parts of the western United States, many experts believe that tropospheric winds from northeastern China are responsible for spreading the KD trigger.
   (a) Once in the upper respiratory tract, the antigen triggers an immune-mediated response (innate and adaptive) within the arterial walls.
   (b) There appears to be a predilection for coronary arteries and other medium-sized muscular arteries with histopathological studies demonstrating necrotizing arteritis, vasculitis, and luminal myofibroblastic proliferation as the three most common vasculopathic processes.
   (c) Depending on the vasculopathic process, the end result is arterial wall weakening leading to aneurysm formation or coronary artery stenosis and thus myocardial ischemia.
2. In addition, it is commonly believed that there is a genetic susceptibility for developing KD.
   (a) This is supported by the fact that Japanese children, regardless of their geographical location, continue to have an increased risk for developing KD.
   (b) Based on the available literature, many experts believe that susceptibility to KD is polygenic and varies among different ethnic groups.
   (c) Epidemiological studies have shown that siblings of children with KD are ten times more likely to develop KD when compared to the general population. Additionally, children of parents with a history of KD are twice as likely to also develop KD.

Incidence

KD usually affects children and has a higher prevalence in Asians with the highest reported annual incidence in Japan of 265 cases per 100,000 children under the age of 5. In the United States, the incidence ranges from 19 to 24.7 per 100,000 with the highest reported annual incidence in California.

Signs

1. Signs of acute infection include fever of 5 days duration, bilateral conjunctivitis, lip and oral mucosal alterations (strawberry tongue, oropharyngeal edema, red fissured lips), polymorphous rash, cervical lymphadenopathy, and

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changes in the extremities such as erythema and edema. The acute phase lasts for 7–14 days followed by a subacute one for 2–4 weeks, when there is desquamation of the hands and feet and thrombocytosis with a risk for coronary artery thrombosis. When all criteria are not met but there is a high clinical suspicion for KD, imaging can be helpful in the diagnosis. Echocardiography and coronary angiography may demonstrate coronary aneurysms, stenosis, or myocardial ischemia.
   (a) The incidence of coronary artery aneurysm formation varies with rates ranging from 15% to 25% being reported in the literature.
   (b) Long-term cohort studies show that 55% of patients who develop coronary artery aneurysms will have regression, with 90% showing regression within 2 years of KD resolution.
2. Sudden death, coronary artery aneurysms, cardiac ischemia, myocardial fibrosis, valvular abnormalities, and heart failure may develop from coronary arteritis. Patients with large aneurysms may develop thrombosis and ischemia. Patients may require stenting, CABG, or transplant. Patients with a history of KD, and these potential complications, are most commonly seen during pregnancy.

Interaction with Pregnancy

1. Studies analyzing the interaction of KD with pregnancy are limited and mainly involve case reports and small case studies. However, the complications of over 80 deliveries in women with a history of KD have been reported.
2. Based on the limited data, there may be an increased rate of premature rupture of membrane and preterm labor though most women deliver at term. There were rare reports of postpartum hemorrhage.
3. Although most mothers do well, there are many reports of cardiac decompensation during pregnancy.
4. The hemodynamic changes during pregnancy may place these women at increased risk for myocardial ischemia during pregnancy as increased heart rate, cardiac output, and contractility all increase myocardial oxygen demand. Additionally, these hemodynamic changes also increase arterial wall stress, which can lead to progression of known aneurysms and potentially aneurysm rupture.

Testing

Case reports have documented the management of parturients presenting with acute KD and parturients with a known history of KD.

1. Parturients with acute KD should have echocardiography to determine wall motion abnormalities, valvulopathies, and aortic root measurements.
   (a) Parturients with acute KD and normal cardiac function can undergo expectant management with systemic therapy for KD involving the use of intravenous immunoglobulin (IVIG).
   (b) Patients with abnormal stress echocardiogram or coronary artery dilation should have further workup with magnetic resonance angiography (MRA).
2. Women with a history of KD with cardiovascular involvement will need stress echocardiography and computed tomography angiography (CTA) prior to pregnancy or MRA after conception to assess the coronary arteries.
3. Women with a known history of KD and an unknown cardiac status should have baseline echocardiography.
   (a) Prior to conception, computed tomography (CT) calcium scores should be obtained. If the latter is abnormal, then either a CTA or MRA is indicated.
   (b) If the patient presents after conception, then a stress echocardiogram and MRA should be performed.
   (c) The need for repeat echocardiograms is dictated based on symptoms or signs of worsening heart failure and/or ischemia.

Management

Medical

1. Management of patients with active KD involves decreasing inflammation and mitigating the development of thrombus and aneurysm formation.
2. Coordinated care between maternal fetal medicine, cardiology, and obstetric anesthesia is imperative.
3. Once the diagnosis of KD is confirmed, treatment with IVIG and aspirin is indicated.
4. If patients are treated with IVIG within the first 10 days of fever onset, the risk of aneurysm formation can be decreased from 25% to 5%. For patients with known aneurysms in whom long-term antiplatelet therapy is indicated, care must be coordinated with maternal fetal medicine.
5. Pregnancy termination should be discussed with patients with severe heart failure or significant ischemia who might benefit from percutaneous coronary interventions or cardiac surgery.

Anesthetic Management

1. Anesthetic management of these patients will vary depending on their preoperative antiplatelet regimen (enoxaparin) and their cardiovascular function/status.

2. The mode of delivery should be dictated by obstetrical indications.
3. When not contraindicated by anticoagulation, neuraxial analgesia and anesthesia may be performed. In fact, labor epidural analgesia will be beneficial to the pregnant woman with myocardial ischemia, aneurysm, and heart failure. Epidural analgesia will decrease maternal tachycardia, hypertension, and oxygen consumption. Assisted second stage is often recommended, and for these patients, epidural analgesia is beneficial.
4. Similarly, a low-dose combined spinal-epidural or epidural could be used for surgical anesthesia. Depending on the severity of cardiac involvement, a spinal may not be the best choice if hypotension and tachycardia could be detrimental.
5. In addition to standard ASA monitors, the need for invasive monitoring (arterial line, central venous pressure, TEE) is indicated on a case-by-case basis. Arrhythmias may occur.
6. The use of intravenous beta-blockers, calcium channel blockers, and nitrates is indicated to closely control blood pressure in patients with known coronary artery aneurysms.
7. Similar to nonpregnant patients at risk for myocardial ischemia, the goals are to maximize myocardial oxygen supply and minimize tachycardia and other determinants of myocardial oxygen demand.

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