Managing Kawasaki Disease

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What is Kawasaki Disease?

Kawasaki Disease is an acute systemic vasculitis of early childhood, with a predilection for involving the coronary arteries. Despite 40 years of research, its aetiology remains unclear.

Epidemiological, clinical and immunological data support the consensus that Kawasaki Disease is due to one or more infectious agent(s), which evoke an abnormal immune response in genetically susceptible individuals.

When should I suspect Kawasaki Disease?

Clinicians should suspect Kawasaki Disease in a child presenting with fever of about five days duration, and with clinical signs such as red eyes, red lips and extremity changes. Other supportive clinical signs include irritability, perineal rash and inflammation (erythema and induration) of the Bacillus Calmette–Guérin (BCG) scar.

How is Kawasaki Disease Diagnosed?

Kawasaki Disease is diagnosed based on the presence of fever and four out of five other clinical criteria (rash, non-purulent conjunctivitis, oropharyngeal changes, lymphadenopathy and changes to the extremities).

The Kawasaki Disease diagnostic criteria share overlapping features with other childhood diseases such as viral fever and drug reaction (e.g. Stevens Johnson Syndrome and Toxic Shock Syndrome). Diagnosis can be difficult, and incomplete Kawasaki Disease, i.e. coronary arterial lesions in the absence of full criteria, is increasingly being recognised.

Other associations include aseptic meningitis, facial nerve paralysis, myocarditis and shock-like syndrome. Unfortunately, there is currently still no diagnostic test available for Kawasaki Disease, but collaborative research is being undertaken by basic scientists and clinicians to identify biomarkers specific to the disease.

What are the Acute Complications of Kawasaki Disease?

Kawasaki Disease is not a benign childhood exanthem. The acute phase of the disease may be associated with inflammation of the coronary arterial wall (Figure 1), myocarditis, pericarditis and valvulitis.

The aim of treating Kawasaki Disease is to prevent its associated cardiac complications. Coronary arterial lesions are the most common, occurring in up to 25 percent of untreated and up to five percent of treated children despite current best treatment.

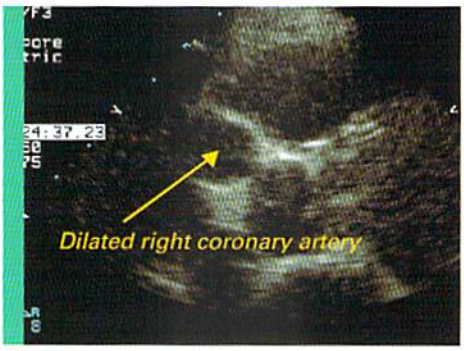

Figure 1: Transthoracic 2D Echocardiogram demonstrating a dilated right coronary artery.

Patients with large coronary arterial dilatations diagnosed via 2D Echocardiography may require other imaging modalities such as cardiac catheter studies (Figure 2), cardiac magnetic resonance imaging and cardiac computed tomography scans to further delineate the extent of coronary arterial involvement.

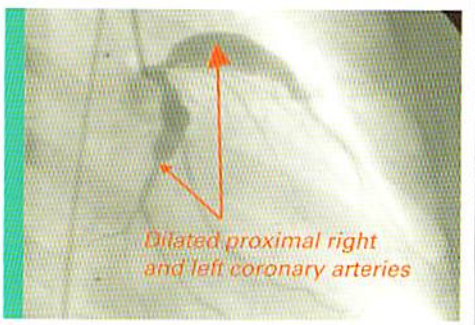

Figure 2: Angiogram showing dilated proximal right and left coronary arteries.

What are the Long Term Complications of Kawasaki Disease?

Coronary arterial aneurysms are the most serious long term complication of Kawasaki Disease. With these lesions, death can occur from rupture of aneurysms and subsequently from myocardial infarction due to coronary arterial stenosis.

The most common paediatric-acquired heart disease in developed countries, Kawasaki Disease currently affects an estimated 32.5 per 100,000 children under five years of age in Singapore.

Case incidence has been noted to be on the rise, with approximately 120 new cases diagnosed and managed at KK Women's and Children's Hospital (KKH) each year.

The risk of sudden cardiac death in patients with Kawasaki Disease is estimated to be two percent of those with coronary arterial lesions. Lifelong medical therapy, coronary arterial grafting and even a heart transplantation may be required.

How is Kawasaki Disease Treated?

Intravenous immunoglobulin (IVIG) is currently the first line of treatment for patients with confirmed Kawasaki Disease. If administered within the first ten days of illness, IVIG lowers the likelihood of coronary arterial aneurysms approximately five-fold.

High doses of aspirin are also started until resolution of fever. The patient is then maintained on low doses of aspirin for six weeks or until the resolution of coronary arterial lesions. For patients who are G6PD deficient, oral dypiridamole may be used in place of asprin.

What are the Areas of Research in Kawasaki Disease?

Extensive research is being conducted on Kawasaki Disease. Basic science research is centered on identifying genetic loci conferring susceptibility to Kawasaki Disease using candidate gene approaches and genome-wide scans (GWAS).

Novel therapeutic agents (e.g. doxycycline) are being studied using murine models. Clinical research includes identification of epidemiological associations, new diagnostic modalities (e.g. myocardial perfusion scanning), biomarker identification and new therapies.