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A REVIEW ON KAWASAKI DISEASE

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ABSTRACT

Kawasaki Disease mostly strikes children under the age of five. Kawasaki Disease is now known to be the leading cause of coronary heart disease in children that affects adult life. It is also suspected to be the primary cause of death by unexplained heart failure in children and young adults. Such cases are on the increase and are believed to be partly hereditary, which poses a 10% chance of being passed onto offspring.^[1]

KEYWORDS: Kawasaki disease, coronary heart disease, children.

INTRODUCTION

Kawasaki Disease (KD) is a form of vasculitis, where blood vessels become inflamed throughout the body. [2] The disease was first described in Japan by Tomisaku Kawasaki in 1967, and the first cases outside of Japan were reported in Hawaii in 1976. KD occurs worldwide, with the highest incidence in Japan, and it most often affects boys and younger children. In the continental United States, population-based and hospitalization studies estimate an incidence of KD ranging from about 9 to 20 per 100,000 children under 5 years of age. In the year 2016, approximately 5440 hospitalizations with KD were reported among children under 18 years of age in the US; 3935 of these children were under 5 years of age, for a hospitalization rate of 19.8 per 100,000 children in that age group. [3]

Background

KD is the second most common vasculitis in childhood after Henoch Schonlein purpura, and is **the most common** cause of acquired heart disease in children in developed countries causing coronary artery aneurysms (CAA). It has a worldwide distribution, although is more common in Asian children. Approximately 85% of cases occur under 5 years of age, peak age 18-24 months. KD in children < 6 months and >5 years is less common, however these children are more likely to develop CAA. [4]

Causes

Scientists haven't found an exact cause for Kawasaki disease. It might be linked to genes, viruses, bacteria, and other things in the world around a child, such as chemicals and irritants. Scientists don't believe the disease is contagious from person to person.^[5]

Risk factors

Three things are known to increase your child's risk of developing Kawasaki disease.

- Age. Children under 5 years old are most at risk of Kawasaki disease.
- **Sex.** Boys are slightly more likely than girls are to develop Kawasaki disease.
- Ethnicity. Children of Asian or Pacific Island descent, such as Japanese or Korean, have higher rates of Kawasaki disease.

Symptoms

Kawasaki disease signs and symptoms usually appear in three phases.

1st phase

Signs and symptoms of the first phase may include:

- A fever that is often is higher than 102.2 F (39 C) and lasts more than three days
- Extremely red eyes without a thick discharge
- Red, dry, cracked lips and an extremely red, swollen tongue
- Swollen, red skin on the palms of the hands and the soles of the feet
- A rash on the main part of the body and in the genital area
- Irritability
- Swollen lymph nodes in the neck and perhaps elsewhere. Cervical, most commonly unilateral, tender. At least one node >1.5cm.

2nd phase

In the second phase of the disease, your child may develop:

 Peeling of the skin on the hands and feet, especially the tips of the fingers and toes, often in large sheets

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- Abdominal pain
- Joint pain
- Diarrhea
- Vomiting

3rd phase

In the third phase of the disease, signs and symptoms slowly go away unless complications develop. It may be as long as eight weeks before energy levels seem normal again. [6]

Investigations

 Echocardiogram – at baseline (this should not delay initiation of treatment) and at 6 weeks. Abnormalities should be managed in consultation with paediatric cardiology and haematology services.

In all patients consider:

- FBE, CRP, ESR, UEC, LFT (NB ESR is unreliable after IVIg administration)
- Blood culture
- · Serum to store
- Urinalysis (sterile pyuria)
- ECG^[7]

Treatment

The goal of therapy in the acute phase is to reduce inflammation and arterial damage and to prevent thrombosis in those with coronary artery abnormalities.

1. Intravenous immunoglobulin (IVIg): 2g/kg as a single IV infusion on diagnosis.

IVIg should always be given within the first 10 days of the illness, but should also be given to patients diagnosed after 10 days of illness if there is evidence of ongoing inflammation.

A second dose of 2g/kg IVIg should be given to patients who do not respond to the first dose, as demonstrated by persistent or recurrent fevers after 36 hours.

- **2. Corticosteroids:** The use of corticosteroids in the treatment of KD remains controversial. Consider for high risk patients in discussion with local paediatric team. High risk as suggested by:
- Signs of shock.
- Patients < 12 months of age.
- Asian ethnicity.
- ALT > 100 IU/L
- Albumin < 30 g/L
- Any patient with evidence of cardiac involvement on echocardiography at time of presentation.

Prednisolone 2mg/kg (max 60mg) orally daily for a minimum of 5 days and until CRP normalizes. Corticosteroids are effective in the treatment of fever in most patients with IVIG-refractory KD. [8]

- 3. Aspirin: 3-5mg/kg as a daily dose until normal echo on follow up (minimum 6 weeks). The association of Reye syndrome with aspirin remains a consideration, thus risks must be balanced against clinical benefit.^[7]
- 4. Plasma exchange: Plasma exchange has been reported in uncontrolled clinical trials to be an effective therapy in patients who are refractory to IVIG and to lower the incidence of coronary artery aneurysms.

Complications

Heart complications include:

- Inflammation of blood vessels, usually the coronary arteries, that supply blood to the heart
- Inflammation of the heart muscle
- Heart valve problems. [10]

Child could develop serious lifelong heart disease after the illness. Coronary aneurysms may develop. [11]

Discharge plan

Patient afebrile for at least 12 hours before discharge-Patient received 6-week supply of low-dose aspirin-Cardiology consult and Echocardiogram completed-Cardiology follow-up and Echocardiogram scheduled in 2 weeks · Family received education materials regarding fever monitoring. [12]

CONCLUSION

Kawasaki disease is an important cause of fever in young children. It results in a high incidence of cardio- vascular damage if not treated promptly. Kawasaki Disease is now known to be the leading cause of coronary heart disease in children that affects adult life.^[1]

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