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Working in the pediatric ED at this time of year is challenging in that it is a constant, 24/7, revolving door of children with fever. As we know, most of these children have viral syndromes of some kind, commonly gastrointestinal or respiratory. Given that we see so many children with this complaint, it can be easy to become complacent in our differential and management and even possibly overlook things. With this in mind, I am going to review Kawasaki Disease, as this is a diagnosis not to be missed.

Clinical Features/Diagnosis:
Kawasaki disease is a **vasculitis of the small and medium sized vessels**, the cause of which still remains unknown. It is most commonly seen in children **under the age of 5** and has a slightly higher prevalence in males compared to females. In typical Kawasaki disease, there are specific clinical diagnostic criteria that can manifest simultaneously or in a series of days. These criteria include **fever (usually >38.5) for greater than or equal to 5 days accompanied by at least 4 of** the following 5 physical exam findings:

-Bilateral, nonexudative conjunctivitis
-Mucositis (classically "strawberry tongue" and/or cracked, red lips)
-Cervical lymphadenopathy
-Peripheral extremity changes
-Polymorphous generalized rash

It should be noted that there are newer current guidelines stating that a child with 5 days of fever and 2 of the above findings should prompt further laboratory testing, as there have been cases of **incomplete** Kawasaki disease, especially in infants. For this reason, an ESR and CRP should be obtained, and if **CRP > 3mg/dl or ESR >40mm/hr**, then further testing should be completed. This includes:

-Albumin </= 3g/dl
-Anemia
-Platelets >/=450,000/mm^3
-WBC >/= 15,000mm^3
-Elevated ALT
-Sterile pyuria

Children with any of the above laboratory abnormalities should be treated empirically while awaiting echo results. Children who do not have lab abnormalities can be monitored daily.

What are we trying to avoid?
Although Kawasaki disease is self-limiting, there are cardiac sequelae that can be devastating. It is for this reason that Kawasaki disease must be recognized and treated promptly. The cardiac complications occur in 2 phases. The first phase usually occurs during the acute febrile period and is characterized by **mild diffuse myocardial inflammation** with tachycardia, nonspecific ST-T wave changes, and a gallop.

Pericardial effusions are also common in this phase, but both usually resolve when fever resolves. The second phase usually occurs 2-4 weeks after onset of illness and is characterized by **coronary artery dilation that can progress to coronary artery aneurysm**. Death in Kawasaki disease is usually due to **myocardial infarction** secondary to coronary occlusion.

How can we prevent sequelae?

In order to prevent poor outcomes with this disease, **supportive therapy combined with aspirin and IVIG** are the mainstays of treatment. IVIG is usually a one-time infusion within the first 7-10 days of illness of 2g/kg over 10-12 hours but can be repeated if there is persistent or recurrent fever. Additionally, aspirin is started at 80-100mg/kg/d divided into an every 6-hour regimen until the child is afebrile for 48-72 hours. The dose is then reduced to 3-5mg/kg/d for 6-8 weeks.

References / Further Reading:

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