PedsCases Podcast Scripts

This is a text version of a podcast from Pedscases.com on "Kawasaki Disease." These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at www.pedcases.com/podcasts.

Kawasaki Disease

Developed by Chris Gerdung and Melanie Lewis for PedsCases.com.
September 5, 2009

Introduction:

Chris: Hi, I'm Chris Gerdung, a medical student from the University of Alberta.

Dr. Lewis: Hi I'm Melanie Lewis, a general pediatrician from the Stollery Children's Hospital in Edmonton Alberta. Today we will be discussing Kawasaki disease.

Chris: Dr. Lewis, I always thought Kawasaki disease was a very rare disease. Why is it so important for students to know?

Dr. Lewis: That's a very good question Chris. In fact, Kawasaki disease is the leading cause of acquired heart disease in children. In my short career, I've already seen at least a dozen cases of typical or atypical Kawasaki disease. And, missing a case' may have severe, life threatening consequences, or even deadly consequences.

The bottom line is, if you see a child who has been febrile for several days, and is extremely irritable, you should at least consider Kawasaki disease as a possibility, before dismissing it as yet another viral illness in your office or emergency department.

Chris: So Dr. Lewis, what is Kawasaki disease then?

Dr. Lewis: Kawasaki disease is an acute inflammatory vasculitis of medium sized arteries. It initially presents with a history of fever in children between the ages of 6 months to *5* years, and after approximately five days of fever, other signs can be observed which point more precisely to the diagnosis of Kawasaki disease. We will be describing the criteria for diagnosing Kawasaki's later in the Podcast.

Etiology:

Chris: Ok, so what causes Kawasaki disease then?

Dr. Lewis: That is a great question, and one that is not entirely known. The current belief is that the disease is caused by an infectious agent in children who are genetically predisposed. This belief is supported by an increase in the number of cases during the

Developed by Chris Gerdung and Melanie Lewis for PedsCases.com.
September 5, 2009

winter and spring seasons, as is commonly seen with other infections. In addition, during epidemics of the disease, the increased numbers of cases spreads in a geographical manner similar to other infectious diseases.

Interestingly, children less than three months of age rarely become infected, which is thought to be due to the protection of the infant by circulating maternal antibodies. In unaffected children, once the maternal antibodies are recycled the child begins to produce their own antibodies in response to antigens that are presented to its immune system. In children who present with Kawasaki disease however, it is thought that a genetic variation does not immediately allow antibodies to be created in response to a ubiquitous virus, resulting in the clinical signs and symptoms seen.

Chris: Can you clarify for me Dr. Lewis; is this a vasculitis or an infectious disease?

Dr. Lewis: Kawasaki disease does involve edema and inflammation of the blood vessels, which is a vasculitis, but the inciting agent is likely to be infectious in nature. The most severely affected blood vessels are the medium sized arteries. Pathologic examination of vessels reveals edema of both endothelial and smooth muscle cells, as well as inflammatory infiltrate into the intima of the arteries. *As* the disease progresses, the elastic properties of the lamina are destroyed resulting in a weakened vessel. This weakening can lead to dilatation and formation of an aneurysm. Additionally, as the inflammatory infiltrate replaces the normal intima, scarring and stenosis can occur. Unfortunately, in Kawasaki disease, the most common complications involve the heart and the coronary arteries.

In 30-50% of cases with Kawasaki disease, coronary artery dilatation begins to occur roughly 10 days after the fever begins, but as early as 8 days. Most dilatation of the coronary arteries is not permanent and the vessel will return to normal size in approximately 6 to 8 weeks, however in some patients the dilation will progress to form an aneurysm. Rarely, aneurysms can continue to dilate to over 8 mm in size, forming giant aneurysms. These aneurysms have a poor prognosis, as they may attempt to heal, causing stenosis of the vessel, which may lead to myocardial infarction. Kawasaki disease *is* the leading cause of acquired heart disease in children. In other patients, the vessel wall may become too thin, and rupture of the aneurysm can occur. Treatment of the disease before the 10th day of fever substantially reduces the risk of these complications from occurring

<u>Diagnosis:</u>

Chris: So, what test is used to diagnose Kawasaki disease then?

Dr. Lewis: Another great question, Chris. Kawasaki disease is actually a clinical diagnosis, and isn't based on one specific diagnostic test. First of all, Kawasaki disease is found in all racial groups, however the incidence is roughly 20 times higher in individuals of Asian background versus Caucasians. In addition, boys and the very young seem to be at the highest risk for coronary artery involvement.

Chris: So if there is no lab test, what are the clinical criteria used to diagnose

Kawasaki disease then?

Dr. Lewis: So, to diagnose Kawasaki disease, the child must have a five-day history of fever, as well as four of the five following clinical signs. The first sign is a rash. It's described as polymorphous exanthema, which is macular or maculopapular that is commonly seen on the trunk and diaper region of the child. It may spread to other parts of the body. Any petechial, vesicular or bullous rash does *not* meet the criteria of polymorphous exanthema and suggests an alternate diagnosis. The second sign is bilateral, bulbar, non-exudative conjunctivitis. The injection typically spares the avascular area around the iris known as the limbus and is not usually painful. And I'll repeat, there should be *no* exudate associated with this conjunctivitis. The third clinical sign is changes to the lips and oral cavity, which can include erythema, drying, cracking, bleeding and peeling.

Strawberry tongue can also be found on exam, however it must be determined if this is a finding caused by scarlet fever, due to a Group A Strep infection, or Kawasaki disease. Oral ulceration and pharyngeal exudates are *not* consistent with Kawasaki disease and point to a different diagnosis. The fourth clinical sign that is used to diagnose Kawasaki disease is changes in the extremities, such as erythema, edema, and induration. Later in the disease, periungual desquamation can be found on both the hands and feet. But I can tell you from clinical experience, if you're seeing this on your first exam, you've missed the boat in terms of a window to actually treat this disease. The fifth characteristic used in the diagnosis is cervical lymphadenopathy, which is often unilateral, and contains at least one lymph node greater than 1.Scm in diameter. It is the least common criteria found on exam, however if present it is usually furn, fixed and can be painful. These signs gradually start progressing over the course of the illness and do not all present at the same time. Therefore, although the diagnosis might not be clear, the clinician must keep Kawasaki disease on the differential diagnosis for any fever that lasts longer than five days. The sooner that Kawasaki disease is recognized, the sooner that treatment can begin, thereby reducing the incidence of serious complications.

While true Kawasaki disease presents with a five day history of fever as well as four out of the five clinical signs that we've discussed, many children will never have all four of these signs. In children with suspected Kawasaki disease who have less than four of the clinical findings, the diagnosis of incomplete or atypical Kawasaki disease is applied.

Chris: Is there any way to tell for sure that incomplete or atypical Kawasaki disease is causing the symptoms in the child?

Dr. Lewis: In order for a case of incomplete or atypical Kawasaki disease to be confirmed, the CRP and ESR must be elevated and the presence of three of the following must be confirmed: Leukocytosis with a high proportion of neutrophils, anemia, thrombocytosis (which is usually present in the third, or second week of the disease), hypoalbuminemia, elevated ALT, and over 10 white blood cells per high power field on urine analysis.

If C-reactive protein and ESR are elevated in a patient with suspected Kawasaki disease and less than three of the laboratory findings are present, an echocardiogram must be

conducted to determine if there are changes to the coronary arteries.

Management:

Chris: So after you've made the diagnosis, what can be done to treat the disease?

Dr. Lewis: Once Kawasaki disease is confirmed, treatment must begin as soon as possible. Treatment of Kawasaki disease should begin within 10 days since the onset of the fever. The first line treatment is 2WK.g of intravenous immunoglobulin. This treatment will not only reduce the fever, but it will also reduce the chance of coronary artery aneurysm. If the fever persists for longer than 36 hours after MG treatment, another dose should be entertained. After the administration of MG, 80mg/Kg/day of aspirin should be given until the fever subsides. At that point, the Aspirin should be reduced to 3-5 mg/Kg/day until a normal echocardiogram is seen at a six-week follow-up appointment.

Chris: Other than the echo that is done at six or eight weeks, is other follow-up necessary for these children?

Dr. Lewis: In terms of follow-up, children with no coronary artery dilatation at the initial presentation, and after six weeks, should receive an echocardiogram every *5* years. For patients with dilatation that resolved by 6 weeks, an echocardiogram should be conducted every 3 to *5* years. For all other patients with coronary artery aneurysm or other cardiac complications, annual follow up with a pediatric cardiologist, including an echo, should be done annually or bi-annually depending on the severity.

Prognosis:

Chris: After treatment, what's the prognosis like for these children?

Dr. Lewis: The prognosis for children with Kawasaki disease varies depending on the presence and severity of complications that arise. For most children, the prognosis is very good. Roughly half of recovered patients with no aneurysm formation have serum lipid abnormalities, however no other complaints. Patients who had aneurysms may still be at risk for ischemic heart disease and myocardial infarction.

Take home points:

Chris: Dr. Lewis, we've talked a lot about Kawasaki disease over the last little while, but is there anything in particular that should take away from this podcast?

Dr. Lewis: Well first of all, Kawasaki disease is not that rare. It may very well walk into your outpatient office or emergency department and it's crucial that you recognize it. So any child who presents with high fevers for a number of days, you *must* consider Kawasaki disease. Without prompt recognition and treatment, a child may face lifelong cardiovascular complications.

Developed by Chris Gerdung and Melanie Lewis for PedsCases.com.
September 5, 2009

Developed by Chris Gerdung and Melanie Lewis for PedsCases.com.
September 5, 2009