www.printo.it/pediatric-rheumatology/IE/intro

Kawasaki Disease

Version of 2016

2. DIAGNOSIS AND THERAPY

2.1 How is it diagnosed?

KD is a clinical or bedside diagnosis. This means that the diagnosis is made only on the basis of a clinical evaluation by a doctor. A definite diagnosis can be made if unexplained high fever lasting for 5 or more days is present along with 4 of the 5 following features: bilateral conjunctivitis (i.e. inflammation of the membrane covering the eyeball), enlarged lymph nodes, skin rash, mouth and tongue involvement and extremities changes. The doctor must verify that there is no evidence of any other disease that could explain the same symptoms. Some children show incomplete forms of the disease, meaning that they present with fewer clinical criteria, making the diagnosis more difficult. Such cases are called incomplete KD.

2.2 How long will the disease last?

KD is an illness with three phases: acute, which includes the first 2 weeks when the fever and the other symptoms are present; sub-acute, from the second to the fourth week, a period in which the platelet count begins to rise and aneurysms can appear; and the recovery phase, from the first to the third month, when all the altered lab tests return to normal and some of the blood vessels abnormalities (such as CAA) are resolved or diminished in size.
If untreated, the disease may run a self-limiting course over about 2 weeks leaving the coronaries damaged all the same.

2.3 What is the importance of tests?

Currently, there is no laboratory test that can help to conclusively diagnose the disease. A cluster of tests, such as elevated ESR (erythrocyte sedimentation rate), high CRP level, leukocytosis (increased number of white blood cells), anaemia (low count of red blood cells), low serum albumin and raised liver enzymes, may help in making the diagnosis. The number of platelets (the cells involved in blood clotting) is generally normal in the first weeks of the disease but begins to rise in the second week, reaching very high counts.
Children should undergo periodic examinations and assessment of blood tests until the platelet counts and ESR return to normal.
An initial electrocardiogram (ECG) and echocardiogram should be performed. The echocardiogram can detect dilatation (widening) or aneurysms by evaluating the shape and size of the coronary arteries. In the case of a child with coronary abnormalities, follow-up echocardiograms and additional studies and evaluations will be needed.

2.4 Can it be treated/cured?

The majority of children with KD can be cured; however, some patients develop heart complications, despite the use of proper treatment. The disease cannot be prevented but the best way to decrease coronary complications is to make an early diagnosis and to start treatment as early as possible.

2.5 What are the treatments?

A child with definite or suspected KD should be admitted to the hospital for observation and monitoring and should be evaluated for possible heart involvement.
To diminish the frequency of heart complications, treatment should be started as soon as the diagnosis is made.
Treatment consists of a single high dose of intravenous immunoglobulin (IVIG) and aspirin. This treatment will diminish the inflammation, dramatically relieving the acute symptoms. High dose IVIG is the essential part of treatment since it is able to reduce the occurrence of coronary abnormalities in a high proportion of patients. Though very expensive, for now it remains the most effective form of treatment. In patients with special risk factors, simultaneous corticosteroids may be

given. Patients who do not respond to one or two doses of IVIG have other therapeutic alternatives, including high dose intravenous corticosteroids and biologic drug therapy.

2.6 Do all children respond to intravenous immunoglobulin?

Fortunately, most children will need just a single dose. Those who do not respond may need a second dose, or doses of corticosteroids. In rare cases, new molecules called biological drugs may be given.

2.7 What are the side effects of drug therapy?

IVIG therapy is usually safe and well-tolerated. Rarely, inflammation of the meninges (aseptic meningitis) can occur.
Following IVIG therapy, live attenuated vaccinations should be postponed. (Discuss each vaccine with your paediatrician). Aspirin at high dose can cause nausea or stomach upset.

2.8 What treatment is recommended after immunoglobulin and high dose aspirin? How long should treatment last?

After the fever settles down (usually in 24-48 hours), the dose of aspirin will be tapered down. The low dose of aspirin is maintained due to its effect on the platelets; this means that the platelets will not stick together. This treatment is useful to prevent formation of thrombi (blood clots) inside the aneurysms or the inner linings of inflamed blood vessels, since thrombus formation inside an aneurysm or blood vessel may lead to cutting off of the blood supply to the areas supplied by the blood vessels (cardiac infarction, the most dangerous complication of KD). Low dose aspirin is continued until normalisation of inflammatory markers and a normal follow-up echo. Children with persistent aneurysms should receive aspirin or other anti-clotting drugs under a doctor's supervision for longer periods.

2.9 My religion does not allow me to use blood and blood products. What about unconventional/complementary therapies?

There is no place for unconventional treatments for this disease. IVIG is

the proven treatment of choice. Corticosteroids might be effective in case IVIG cannot be used.

2.10 Who is involved in the medical care of the child?

The paediatrician, the paediatric cardiologist and the paediatric rheumatologist may take care of the acute stage and follow-up of children with KD. In places where a paediatric rheumatologist is not available, the paediatrician along with the cardiologist should monitor patients, especially the ones who have had heart involvement.

2.11 What is the long-term evolution (prognosis) of the disease?

For the majority of patients, the prognosis is excellent, as they will develop a normal life, with normal growth and development.
The prognosis of patients with persistent coronary arteries abnormalities depends mainly on the development of vascular narrowing (stenosis) and obstructions (occlusions). They may be prone to heart symptoms in early life and may need to stay under the care of a cardiologist experienced in the long-term care of children with KD.