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Rheumatic Fever And Post-streptococcal Reactive Arthritis

Version of 2016

1. WHAT IS RHEUMATIC FEVER

1.1 What is it?

Rheumatic fever is a disease caused by throat infection by a bacterium called streptococcus. There are several groups of streptococci but only group A causes rheumatic fever. Although streptococcal infection is a very common cause of pharyngitis (throat infection) in school-age children, not all children with pharyngitis will develop rheumatic fever. The disease may cause inflammation and damage to the heart; it presents first with short-lived joint pains and swelling, and later with carditis (inflammation of the heart) or an abnormal involuntary movement disorder (chorea) due to brain inflammation. Skin rashes or skin nodules may also occur.

1.2 How common is it?

Before antibiotic treatment became available, the number of cases was high in countries with warm climates. After antibiotic treatment became a common practice for treating pharyngitis, the frequency of this disease decreased, but it still affects many children from 5-15 years of age all over the world, leading to heart disease in a small proportion of cases. Because of its joint manifestations, it is included among the many rheumatic diseases of children and adolescents. The burden of rheumatic fever is unequally distributed around the world.
The occurrence of rheumatic fever varies from country to country: there are countries where no case is registered and countries with medium or

high rates (more than 40 cases per 100,000 persons per year). It is estimated that there are over 15 million cases of rheumatic heart disease worldwide, with 282,000 new cases and 233,000 deaths annually.

1.3 What are the causes of the disease?

The disease is the result of an abnormal immune response to throat infection with Streptococcus pyogenes, or Group A β haemolytic Streptococcus. Sore throat precedes the disease onset by an asymptomatic period of variable length.
Antibiotic treatment is needed to treat the throat infection, stop stimulation of the immune system and prevent new infections, because new infections can cause a new disease attack. The risk of a repeated attack is higher in the first 3 years after disease onset.

1.4 Is it inherited?

Rheumatic fever is not a hereditary disease, since it cannot be transmitted directly from parents to their children. However, there are families with several members who developed rheumatic fever. This may be due to genetic factors associated with the possibility of transmission of streptococcus infections from person to person. Streptococcal infection is transmissible through respiratory airways and saliva.

1.5 Why does my child have this disease? Can it be prevented?

The environment and the streptococcus strain are important factors for the development of the disease, but in practice it is difficult to predict who will get it. Arthritis and heart inflammation are caused by abnormal immune response against streptococcus proteins. The chances of getting the disease are higher if certain types of streptococcus infect a susceptible person. Crowding is an important environmental factor, since it favours the transmission of infections. Prevention of rheumatic fever relies on prompt diagnosis and antibiotic treatment (the recommended antibiotic is penicillin) of streptococcal throat infection in healthy children.

1.6 Is it infectious?

Rheumatic fever by itself is not infectious, but streptococcal pharyngitis is. Streptococci are spread from person to person and therefore infection is associated with crowding at home, in schools or at the gym. Careful hand washing and avoidance of close contact with individuals with streptococcal throat infections is important to stop the spread of the disease.

1.7 What are the main symptoms?

Rheumatic fever usually presents a combination of symptoms that may be unique in each patient. It follows streptococcal pharyngitis or tonsillitis not treated with antibiotics.
Pharyngitis or tonsillitis can be recognized by fever, sore throat, headache, red palate and tonsils with purulent secretions and enlarged and painful neck lymph nodes. However, these symptoms can be very mild or completely absent in school-age children and adolescents. After the acute infection has resolved, there is an asymptomatic period of 2-3 weeks. Then the child may present with fever and the signs of the disease described below.

Arthritis

Arthritis most commonly affects several large joints at the same time or may move from one joint to another affecting one or two at a time (knees, elbows, ankles or shoulders). It is called "migratory and transient arthritis". Arthritis affecting hands and cervical spine is less common. Joint pain may be severe even when swelling is absent. Note that pain usually subsides promptly after administration of anti-inflammatory drugs. Aspirin is the most used anti-inflammatory drug.

Carditis

Carditis (heart inflammation) is the most serious manifestation. Accelerated heartbeat during rest or sleep can elicit the suspicion of rheumatic carditis. Abnormal heart examination, with the presence of heart murmurs, is the main sign of heart involvement. It varies from a subtle to a loud murmur that may indicate inflammation of the heart valves, known as "endocarditis". If inflammation is located in the heart sac, known as "pericarditis", some fluid may collect around the heart

but this usually causes no symptoms and clears on its own. In the most severe cases of myocarditis, the heart may become inflamed and pumping may be weakened. It can be recognized by cough, chest pain and accelerated pulse and breathing. Referral to a cardiologist and tests may be indicated. Rheumatic valvular heart disease may be a result of the first attack of rheumatic fever, but it is usually a consequence of repeated episodes and might become a problem later in adult life, so prevention is crucial.

Chorea

The term chorea is derived from a Greek word meaning dance. Chorea is a movement disorder resulting from inflammation of parts of the brain controlling the coordination of movements. It affects 10-30% of patients with rheumatic fever. Unlike arthritis and carditis, chorea appears later during disease course, from 1 to 6 months after the throat infection. Early signs are poor handwriting in school age patients, difficulties with dressing and self-care, or even with walking and feeding, due to trembling involuntary movements. Movements may be suppressed voluntarily for short periods, may disappear during sleep or be exacerbated by stress or fatigue. In students, it reflects on academic achievements due to poor concentration, anxiety and mood instability with easy crying. If subtle, it might be overlooked as a behavioural disturbance. It is self-limited, although supporting treatment and follow-up is needed.

Skin rashes

Less common manifestations of rheumatic fever are the skin rashes called "erythema marginatum", which look like red rings, and "subcutaneous nodules" that are painless mobile grain nodules with normal overlying skin colour, usually seen over the joints. These signs are present in less than 5% of cases and may be overlooked because of their subtle and transient appearance. These signs are not isolated, but they occur together with myocarditis (inflammation of the heart muscle). There are other complaints that may be first noticed by parents such as fever, fatigue, loss of appetite, pallor, abdominal pain and nosebleeds, which may occur in early stages of the disease.

1.8 Is the disease the same in every child?

The most common presentation is the heart murmur in older children or adolescents with arthritis and fever. Younger patients tend to present with carditis and less severe joint complaints.
Chorea may present itself as the only manifestation or may be combined with carditis, but close follow-up and examination by a cardiologist are recommended.

1.9 Is the disease in children different from the disease in adults?

Rheumatic fever is a disease of school children and young people up to 25 years of age. It is rare before the age of 3 and more than 80% of patients are between 5 and 19 years old. However, it may occur later in life if there is non-compliance with permanent antibiotic prevention.