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ORIGINAL PAPER

Rheumatic Heart Disease: Causes, Symptoms, and Treatments

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Abstract Rheumatic heart disease is a major problem in developing countries and is the cause of most of the cardiovascular mortality in young people, leading to about 250,000 deaths per year worldwide. This disease results from an abnormal autoimmune response to group A streptococci infection in a genetically susceptible host. The acute rheumatic fever, the precursor to rheumatic heart disease, can affect various organs and lead to irreversible valve damage and heart failure. The antibiotic Penicillin is generally used for the treatment and prevention of this disease. The treatment guidelines have emphasized antibiotic prophylaxis against recurrent episodes of acute rheumatic fever which is feasible and cost effective. The early diagnosis and treatment is possible if people at risk for rheumatic heart disease in endemic areas are screened. An active surveillance with echocardiography based screening may be a useful program (Marijon et al., Lancet 379(9891):953–964, 2012). The use of vitamins, C and E, and natural antioxidants protect many heart diseases.

Keywords Rheumatic heart disease · Antibiotics · Antioxidants · Autoimmune response

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Introduction

The cardiovascular diseases (CVD) include heart, blood vessels, and blood circulation diseases which are leading cause of death in adults. There are six main types of CVD: the Ischemic heart disease, the lack of oxygen supply; the cerebro-vascular disease, stroke, blood circulation problem in the brain; myocardial infarction (MI), heart attack due to plaques formation in coronary arteries of heart; perivascular disease, blood circulation problem of pain in legs when walking; rheumatic heart disease, due to streptococcal bacterial infection; and congenital heart disease, may be a hole in inside wall of the heart, which is a birth defect from childhood. The rheumatic heart disease is one of the most common heart diseases in Canada, and in many underdeveloped countries of the world [10]. Approximately 250,000 heart patients die of rheumatic heart disease every year in the world [7]. Rheumatic heart disease begins with a bacterial infection in childhood which affects joints and heart valves. In this article the causes, diagnosis, symptoms, and treatments of rheumatic heart disease are mentioned.

Causes and Facts of Rheumatic Heart Disease

The World Heart Federation [13] described rheumatic heart disease (RHD) as one of the most common acquired disease among children of many countries particularly the poor ones. The RHD is caused by rheumatic fever which is due to group A streptococci infection in throat, lungs, and heart, and this fever can be controlled by antibiotics such as Penicillin. The regular monthly injections of antibiotics can prevent rheumatic fever and protect heart valves from further damage. Acute RHD affects heart joints and central nervous system (CNS). The rheumatic fever can cause fibrosis of heart valves which leads to crippling valvular

heart disease and heart failure or death of the patient. The lack of antibiotics availability and more sophisticated drugs in underdeveloped nations as well as poor living conditions and sanitation problems results into Streptococcal infections. The overcrowding, poor housing conditions, under or malnutrition, and lack of access to healthcare all play a major role in the persistence of RHD in developing countries. About 15.6 million people are estimated to be currently affected by RHD with a significant number of them requiring repeated hospitalization, and an unaffordable heart treatment or surgery that may cause several patients to die within 5–20 years [13]. The worst-affected areas are sub-Saharan Africa, South-Central Asia, the Pacific and indigenous populations of Australia, and New Zealand. Up to 1 % of all school children in Africa, Asia, Eastern Mediterranean region, and Latin America show signs of the rheumatic heart disease.

Diagnosis of Rheumatic Heart Disease

The rheumatic heart disease can be diagnosed for the evidence of Streptococcal infection, an increase in antistreptolysin O titer or Deoxyribonuclease [1, 4, 6, 9, 11]. The echocardiographic and Doppler (E&D) studies have identified subclinical Carditis in patients with acute rheumatic fever which suggested RHD in heart patients who had isolated cases of Sydenham chorea [5].

Symptoms of Rheumatic Heart Disease

There are several symptoms of rheumatic heart disease such as rheumatic fever, and many major and minor criteria [1, 3] which are as follows:

The major criteria are

- *Polyarthritis* A temporary migrating inflammation of the large joints, usually starting in the legs and migrating upwards.
- *Carditis* Inflammation of the heart muscle (Myocarditis) which can manifest as congestive heart failure with shortness of breath, Pericarditis with a rub or a new heart murmur.
- *Subcutaneous nodules* Painless, firm collections of collagen fibers over bones or tendons. They commonly appear on the back of the wrist, the outside elbow, and the front of the knees.
- *Erythema marginatum* A long-lasting reddish rash that begins on the trunk or arms as macules, which spreads outward and clear in the middle to form rings, and continues to spread and coalesce with other rings, ultimately taking on a snake-like appearance. This rash typically spares the face and gets worse with heat.
- *Sydenham's chorea (St. Vitus' dance)* A characteristic series of rapid movements without purpose of the face and arms. This usually occurs very late in the disease for at least 3 months from onset of infection.

The minor criteria are

- Rheumatic fever of 38.2–38.9 °C (100.8–102.0 °F),
- *Arthralgia* Joint pain without swelling (only if polyarthritis is not present as a major symptom),
- Raised erythrocyte sedimentation rate or C-reactive protein,
- Leukocytosis,
- ECG showing features of heart block, such as a prolonged PR interval [1], Boon (Only when carditis is not present as a major symptom),
- Previous episode of rheumatic fever or inactive heart disease.

The Other Signs and Symptoms

- Abdominal or stomach pain
- Bleeding from nose
- Preceding Streptococcal infection: Recent Scarlet fever, raised Antistreptolysin O or other Streptococcal antibody titer, or positive throat bacteria culture [1].

Treatment of Rheumatic Heart Disease

Primary prevention of acute rheumatic fever (the prevention of initial attack) is achieved by treatment of acute throat infections caused by group A Streptococcus bacteria. This is achieved by up to 10 days of an oral antibiotic (usually Penicillin) or a single injection of Penicillin. The people who have previous attack of rheumatic fever are at a greater risk of further attacks which can damage the heart. In case of acute rheumatic heart disease the antibiotics are given for several years to prevent fever and heart disease. The open heart surgery is often required to repair or replace heart valves in patients with severely damaged heart valves, the cost of which is very high and poor patients cannot afford the treatment.

The acute rheumatic fever reduction of inflammation is performed with antiinflammatory medications such as Aspirin or corticosteroids. People with positive cultures for Streptococci bacteria in their throat should also be treated with antibiotics. Aspirin is a drug of choice and should be given at high dose of 100 mg/kg/day. But patients should watch for the side effects like gastritis and salicylate poisoning. In some children the use of Aspirin and salicylic acid can be associated with Reye's syndrome which is potentially a deadly condition. The alternative treatment must be considered when administering Aspirin in children and young

people. Ibuprofen for pain and discomfort and corticosteroids for moderate to severe inflammatory reactions should be given for rheumatic fever in children and adolescents. The use of steroids may prevent further scarring of tissue and may prevent development of sequelae such as mitral stenosis. Monthly injections of antibiotic, such as Penicillin must be given for a period of 5 years in patients having one attack of rheumatic fever. If carditis is also present, the length of therapy could be up to 40 years. The treatment of rheumatic fever also includes the continual use of low doses of antibiotics (such as Penicillin, Sulfadiazine or Erythromycin) to prevent re-occurrence of RHD.

Use of Vaccine

Currently, there are no vaccines available to protect against streptococcus pyogenes infection, although research is in progress for the development of a vaccine. There are some difficulties in developing a vaccine for RHD because a wide variety of bacterial strains of streptococcus pyogenes are present in the environment and the large sum of resources such as time and persons will be required for appropriate trials to find out the safety and efficacy of RHD vaccine [12].

Infection Control

The RHD patients with positive cultures for streptococcus pyogenes should be treated with antibiotic penicillin as long as there is no allergy to this antibiotic. For the treatment of acute RHD, Benzathine benzylpenicillin could also be used as mentioned in Oxford Handbook of Clinical Medicine for rheumatic fever.

Inflammation and Pain

Patients with the symptoms of inflammation and pain may require drugs such as corticosteroids and salicylates which are useful for pain control.

Congestive Heart Failure

If heart patients develop significant Carditis which manifests as congestive heart failure, they require usual treatment for heart, the ACE inhibitors, diuretics, beta-blockers, and Digoxin. Unlike normal heart failure, rheumatic heart failure responds well to corticosteroids.

Conclusion

As reported in Rheumatic Heart Disease Information [2], RHD is the most serious complication of rheumatic fever and chronic rheumatic heart disease. About 30 million children and young adults in underdeveloped countries suffer from this heart disease. To prevent rheumatic fever Aspirin and Corticosteroids are used as antiinflammatory medications. The antibiotics (such as penicillin, sulfadiazine, and erythromycin) are used as treatment to control the bacteria, Streptococcus pyogenes infection. At present there is no cure of rheumatic heart disease. NIH, USA [8] reported that RHD develops in approximately 3 % of untreated Streptococcus throat infections. The symptoms of rheumatic fever, irregular heartbeat, nodes under the skin, physical pain in joints, and fatigue are observed. The use of vitamins C, vitamin E, Sytrinol, Pantothenic acid, Niacin, Policosanol, and antioxidants reduce the damage to the heart muscle in rheumatic fever. The natural antioxidants which are found abundantly in the fruits and vegetables such as, apples, blueberries, broccoli, cherries, cranberries, grapes, and spinach all protect from the heart diseases. The life-style changes such as no smoking, diet low in salt, sugar, and fat as well as maintaining body weight and daily physical exercise can prevent several heart diseases.

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