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Review Article

Rheumatic Heart Disease

Tripti Kumar[1], Mohit Singh[2]

[1,2]Sarvepalli Radhakrishnan University Bhopal, India.

I N F O

Corresponding Author:
Mohit Singh, Sarvepalli Radhakrishnan University Bhopal, India.
E-mail Id:
mohit88singh@gmail.com
Orcid Id:
https://orcid.org/0009-0004-7276-6926
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A B S T R A C T

Rheumatic heart disease (RHD) is a chronic heart condition that develops as a complication of untreated or inadequately treated streptococcal throat infections, particularly those caused by the Group A streptococcus bacteria. Although once a significant global health concern, improvements in healthcare and antibiotics have reduced its prevalence in many developed countries. However, RHD remains a serious issue in low-resource settings, affecting vulnerable populations, especially children and young adults.

Keywords:

Introduction

Rheumatic Heart Disease (RHD) is the most commonly acquired cardiovascular disease in children. It is more common in developing countries. RHD is basically a common sore throat. It is caused by Group A beta haemolytic streptococci bacteria. If neglected, it can lead to a severe heart condition. It is caused by rheumatic fever (RF). This is an inflammatory disease. It affects a wide range of connective tissues. In most cases, it affects the heart, but it can also affect the joints, the skin or the brain. The heart valves can become inflamed and scarred over time.[1]

Epidemiology

While the incidence of RHD has significantly declined in high-income countries, it remains prevalent in parts of sub-Saharan Africa, South Asia, the Pacific, and Indigenous populations. RHD is a major public health problem in developing countries, and in low-income communities. Worldwide, it is estimated that about 30 million people suffer from RHD. As of 2015, an estimated 30,000 deaths and 11,500,000 disability-adjusted life years have been attributed to RHD. In India, RHD is most common in the age group of 5–15 years and is estimated to affect between 5–7% of the population. It accounts for between 20% and 30% of hospital admissions related to cardiovascular diseases.

Pathogenesis

Acute rheumatoid fever is an autoimmune reaction of the body to infections caused by Group A beta haemolytic streptococci. One of the reasons for this is that the molecules of the bacteria are similar to those of the host tissue. When the body creates antibodies against these molecules, the host tissues that contain similar molecules are also damaged. Another theory is that the bacteria's antigens get into the blood vessel walls and interact with a few proteins to create a new protein. Since the host's immune system is unfamiliar with this new protein, it perceives it as foreign and forms antibodies against it. The host proteins that contain similar neo-antigens are also damaged.[2]

Risk Factors

- Low socio-economic status
- Over-crowding
- Age- most common in children between the age of 5–15 years

- Climate and season - more in the rainy season and cold climate
- Previous history of RF
- Genetic predisposition[2]

Pathophysiology

The pathophysiology of RHD involves a cascade of inflammatory events that lead to cardiac tissue damage, primarily affecting the heart valves.

1. Initiation of Immune Response

The process begins with the invasion of the throat by Group A streptococcal bacteria. In susceptible individuals, the bacteria stimulate the immune system to mount a defence. During this phase, the immune system produces antibodies to target and eliminate the streptococcal infection.

2. Autoimmune Reaction

In some cases, the antibodies produced to combat the streptococcal infection cross-react with antigens present on the surface of cardiac tissues, particularly heart valves. This phenomenon is known as molecular mimicry, where the immune system, in its attempt to clear the infection, mistakenly identifies cardiac tissues as foreign invaders.

3. Inflammatory Response

The autoimmune reaction triggers an inflammatory response within the heart. Inflammatory mediators, such as cytokines and immune cells, infiltrate the cardiac tissues, causing swelling, redness, and cellular damage. This inflammatory cascade, if left unchecked, can lead to the formation of nodules called Aschoff bodies.

4. Aschoff Bodies and Carditis

Aschoff bodies are characteristic pathological structures seen in the heart tissues during the acute phase of RF. These nodules contain activated immune cells and fibrinoid material. The inflammation, especially in the heart's muscular and valve tissues (carditis), contributes to the structural alterations that are hallmarks of RHD.

5. Valve Involvement

Heart valves, particularly the mitral and aortic valves, are commonly affected in RHD. The inflammatory process causes thickening, scarring, and fusion of the valve leaflets. This can result in stenosis (narrowing) or regurgitation (leaking) of the valves, compromising their ability to regulate blood flow effectively.

6. Chronic Fibrosis and Calcification

Over time, the acute inflammation transforms into a chronic state characterised by fibrosis and calcification of the affected valve tissues. Fibrous adhesions may form between valve leaflets, further impairing their movement. Calcification contributes to the loss of valve elasticity and functionality.

7. Haemodynamic Consequences

The structural changes in the heart valves have profound haemodynamic consequences. Valve stenosis impedes the forward flow of blood, leading to increased pressure within the heart chambers. Regurgitation causes blood to flow backwards, resulting in volume overload and increased workload on the heart.

8. Ventricular Hypertrophy and Heart Failure

The chronic pressure and volume overload place strain on the heart muscle, leading to ventricular hypertrophy, and enlargement of the heart chambers. Over time, this hypertrophy may progress to heart failure, a condition where the heart is unable to pump blood effectively, leading to a myriad of systemic complications.

9. Recurrent Episodes and Progressive Damage

Individuals with a history of acute RF are at an increased risk of recurrent episodes, further exacerbating cardiac damage. Each recurrence contributes to cumulative and progressive valvular damage, perpetuating the cycle of inflammation, fibrosis, and calcification.[3]

Clinical Features

RHD is always caused by RF and Streptococcal Infection. Therefore, a history of RF or Streptococcal infection in the last few years is essential for diagnosing RHD. RF has many characteristics and symptoms usually start after about 1 to 5 weeks after an episode of Step Throat. In many cases, the throat infection is mild and missed, or the child has recovered from the infection before going to the doctor.

RF can come with any of the following symptoms:

- Fever
- Joints that are swollen, tender, and red and very painful (especially the knees and ankles)
- Nodules (small bumps under the skin)
- Red, raised rash that looks like a lattice-like structure, usually on your chest, back, or abdomen
- Chest discomfort and shortness of breath
- Uncontrolled movements of hands, feet, or face muscles
- Weakness

The symptoms of RHD vary depending on the extent of damage to the heart valves. The most common signs and symptoms are shortness of breath, chest pain, and swelling.

RF, triggered by an untreated streptococcal infection, leads to an autoimmune response targeting various tissues,

particularly heart valves. Chronic inflammation and scarring result in valvular damage, leading to the development of RHD.[2,3]

Clinical Manifestations

Cardiac Symptoms

- **Murmurs:** One of the hallmark clinical features of RHD is the development of heart murmurs, primarily due to valvular involvement. Mitral and aortic valves are most commonly affected.
- **Dyspnea:** As the disease progresses, patients may experience shortness of breath, especially on exertion, reflecting the impaired function of affected heart valves.
- **Chest Pain:** Patients with RHD may complain of chest pain, often associated with exertion or related to heart failure.[4]

Systemic Involvement

- **Arthritis:** Acute RF often presents with migratory arthritis, affecting multiple joints. This symptom is a key diagnostic criterion for RF.
- **Subcutaneous Nodules:** In some cases, subcutaneous nodules may develop, usually over bony prominences, adding to the diagnostic criteria of RF.
- **Erythema Marginatum:** A rare but distinctive skin manifestation, erythema marginatum, may occur, presenting as non-pruritic, pink, or erythematous lesions with well-defined borders.

Central Nervous System Involvement

Sydenham's Chorea: Neurological involvement may manifest as Sydenham's chorea, a movement disorder characterised by rapid, purposeless, and involuntary movements.[5]

Prevention

Prevention can be done at three levels:

a) Primordial

b) Primary

c) Secondary

Primordial Prevention: At this level of prevention, prevention includes the prevention of the risk factors for the disease. If there are no risk factors, there is no risk of attacks of Streptococcal Pharyngitis and no risk of Pharyngitis resulting in RHD. As mentioned earlier, the risk factors for RHD are poor socioeconomic status, overcrowding, malnourishment etc. Therefore, the steps of primary prevention would be - governmental measures to improve environmental conditions and provide adequate means and access to healthcare.

Primary Prevention: This level of prevention applies when the streptococcal episodes of pharyngitis are already present. Therefore, the goal of this level of prevention would be to avoid the development of RF in the patient. This can be accomplished through effective treatment and elimination of the infection through antibiotic therapy.

Penicillin is the drug used for treatment. It is preferred over other drugs because it is less expensive and there is less evidence of penicillin-resistant bacteria.

A 10-day oral treatment schedule is used. If it appears that the patient will not be able to tolerate the medication, a single intravenous (IV) injection can be administered containing benzathine benzoquinoline penicillin.

Erythromycin can be used to treat penicillin allergy.[5]

Secondary Prevention: Secondary prevention of RF is defined as the continuous administration of specific antibiotics to patients with previous attacks of RF or a well-documented RHD. The purpose is to prevent colonisation or infection of the upper respiratory tract with Group A beta haemolytic streptococci and the development of recurrent attacks of RF. Secondary prophylaxis is mandatory for all patients who have had an attack of RF, whether or not they have residual rheumatic valvular heart disease.[5]

- Secondary prevention is done by giving long-acting benzathine penicillin.
- Dose – 1.2 million units every 3 weeks or 0.6 million units every alternate week.

Duration of prophylaxis:

- Patient without carditis – till 5 years after their last episode or till the age of 18 years (whichever is of longer duration).
- Patient with carditis – till 10 years after their last episode or till the age of 25 years (whichever is of longer duration).
- Patients with established RHD should receive lifelong treatment.[6]

Conclusion

RHD is a poverty-driven condition that begins with a sore throat and causes long-term damage to the heart tissue. With good health education, communication, and basic prevention, this condition can be largely controlled. The importance of this condition is being re-evaluated around the world, and several research studies are currently underway to improve prevention and treatment plans for rheumatoid fever and other forms of heart disease.

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References

1. Watkins DA, Beaton AZ, Carapetis JR, Karthikeyan G, Mayosi BM, Wyber R, Yacoub MH, Zühlke LJ. Rheumatic heart disease worldwide: JACC scientific expert panel. J Am Coll Cardiol. 2018 Sep 18;72(12):1397-416. [PubMed] [Google Scholar]
2. Remenyi B, Carapetis J, Wyber R, Taubert K, Mayosi BM. Position statement of the World Heart Federation on the prevention and control of rheumatic heart disease. Nat Rev Cardiol. 2013 May;10(5):284-92. [PubMed] [Google Scholar]
3. Katzenellenbogen JM, Ralph AP, Wyber R, Carapetis JR. Rheumatic heart disease: infectious disease origin, chronic care approach. BMC Health Serv Res. 2017 Nov 29;17(1):793. [PubMed] [Google Scholar]
4. Cannon J, Roberts K, Milne C, Carapetis JR. Rheumatic heart disease severity, progression and outcomes: a multi-state model. J Am Heart Assoc. 2017 Mar 2;6(3):e003498. [PubMed] [Google Scholar]
5. Marijon E, Mirabel M, Celermajer DS, Jouven X. Rheumatic heart disease. Lancet. 2012 Mar 10;379(9819):953-64. [PubMed] [Google Scholar]
6. Carapetis JR, Beaton A, Cunningham MW, Guilherme L, Karthikeyan G, Mayosi BM, Sable C, Steer A, Wilson N, Wyber R, Zühlke L. Acute rheumatic fever and rheumatic heart disease. Nat Rev Dis Primers. 2016 Jan 14:2:15084. [PubMed] [Google Scholar]