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# Rheumatic Heart Disease

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## Continuing Education Activity

Rheumatic heart disease is a systemic immune condition that occurs as a complication of rheumatic fever. This occurs after a beta-hemolytic streptococcal infection of the throat. Rheumatic heart disease is a critical form of acquired heart disease in children and adults worldwide. This activity illustrates the evaluation and management of rheumatic heart disease and reviews the role of the interprofessional team in improving care for patients with this condition.

## Objectives:

- Identify the aberrant immune response to *Streptococcus pyogenes* infection in the pathophysiology of rheumatic heart disease.
- Outline the patient history associated with rheumatic heart disease.
- Summarize the echocardiographic criteria for the diagnosis of rheumatic heart disease.
- Explain the importance of improving care coordination amongst the interprofessional team to stress the importance of strict follow-up to improve outcomes in those with rheumatic heart disease.

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## Introduction

Rheumatic heart disease is a systemic immune process that is sequelae to a beta-hemolytic streptococcal infection of the pharynx. It is most common in developing countries. It is responsible for 250,000 deaths in young people worldwide each year. Over 15 million people have evidence of rheumatic heart disease.[1]

## Etiology

Rheumatic heart disease results from either a single or repeated attacks of rheumatic fever that results in rigidity and deformity of valve cusps, the fusion of the commissures, or shortening and fusion of the chordae tendineae. Over 2 to 3 decades, valvular stenosis and/or regurgitation results. In chronic rheumatic heart disease, the mitral valve alone is the most commonly affected valve in an estimated 50% to 60% of cases. Combined lesions of both the aortic and mitral valves occur in 20% of cases. Involvement of the tricuspid valve occurs in about 10% of cases but only in association with mitral or aortic disease. Tricuspid valve cases are thought to be more common when recurrent infections have occurred. The pulmonary valve is rarely affected.

## Epidemiology

Rheumatic heart disease (RHD) is the most critical form of acquired heart disease in children and young adults living

in developing countries. RHD accounts for approximately 15 to 20 percent of all patients with heart failure in endemic countries.[2]

A study of rheumatic heart disease cases estimated that in 2015, there were globally 33.4 million cases of RHD, 10.5 million disability-adjusted life-years due to RHD, and 319400 deaths due to RHD.[3] The incidence of rheumatic heart disease is highest in Oceania, central sub-Saharan Africa, and South Asia. In 2015, there were noted to be 3.4 cases per 100,000 population in nonendemic countries and 444 cases per 100,000 population in endemic countries.[3]

Rheumatic heart disease affects predominantly those living in poverty with inadequate access to health care and unchecked exposure to group A streptococcus. A systematic review and meta-analysis calculated the prevalence of clinically silent RHD (21.1 per 1000 people) to be approximately seven to eight times higher than that of clinically manifest disease (2.7 per 1000 people). The prevalence of rheumatic heart disease increases with age, from 4.7 per 1000 children at 5 years of age to 21.0 per 1000 children at 16 years of age.[4] Based on this data, estimates are that the RHD burden could increase by as much as double than that in the Global Burden of Disease study. Based on the fact that children in sub-Saharan Africa represent 6 to 7 percent of the total global RHD burden, there may be an estimated 50 to 80 million persons currently affected with RHD worldwide.[5]

## Pathophysiology

Rheumatic heart disease is the result of valvular damage caused by an abnormal immune response to *Streptococcus pyogenes* infection, which is classified as a group A streptococcus that causes acute rheumatic fever.[6]. Acute rheumatic fever occurs around three weeks after group A streptococcal pharyngitis and can affect joints, skin, brain, and heart.[7] After multiple episodes of rheumatic fever, progressive fibrosis of heart valves can occur, which can lead to rheumatic valvular heart disease. If valvular heart disease remains untreated, then heart failure or death may occur. The precise pathophysiology is not well known.[6] Rheumatic carditis is characterized by Aschoff nodules and MacCallum plaques. Aschoff bodies are seen in nodules in the hearts affected with rheumatic fever. They result from inflammation in the heart muscle. MacCallum plaques are seen on the valves and the subendocardium in the left atrium.

## History and Physical

Rheumatic fever is the primary cause of acquired heart disease in children and young adults worldwide. Rheumatic fever occurs 2 to 3 weeks after a group A beta-hemolytic streptococcal pharyngeal infection.[8]

Carditis is the most serious presentation of rheumatic fever. The symptoms and signs of carditis are dependent on the areas of the heart involved, which include the pericardium, myocardium, or heart valves. The presentation of a pericardial friction rub on auscultation leans toward the diagnosis of pericarditis. The presence of signs of congestive heart failure points toward a diagnosis of myocarditis, which includes but is not limited to lower extremity edema, shortness of breath with exertion or rest, abdominal distension, or inability to lay flat due to shortness of breath (orthopnea). Myocarditis in the absence of valvular disease is unlikely to be rheumatic in origin. Therefore, an apical systolic or basal diastolic murmur should be auscultated on physical exam. Mitral regurgitation is the most common valvular lesion, which is an apical pan-systolic murmur on auscultation.[8] Aortic regurgitation is less common. If patients have a known history of rheumatic heart disease, a change in the character of the murmur or the presence of a new murmur on auscultation leads to the diagnosis of acute rheumatic heart fever. Rheumatic heart disease predominantly affects the left-sided cardiac valves.[8] The tricuspid valve and rarely pulmonary valve can be affected, but very unlikely without mitral valve involvement.

## Evaluation

Rheumatic heart disease has a variety of clinical manifestations including myocarditis, decompensated congestive heart failure, arrhythmias (i.e., atrial fibrillation), and valvular heart disease.

Myocarditis can result in conduction disturbances in the heart. Therefore, an electrocardiogram (EKG) is necessary. An EKG can show varying forms of heart block including first-degree, second-degree, or third-degree AV block.

A chest x-ray should be completed to evaluate for cardiomegaly or pulmonary vascular congestion, which can be signs of congestive heart failure.

A transthoracic echocardiogram is more sensitive and specific than auscultation during the physical examination for detection of rheumatic heart disease. Rheumatic heart disease seen on transthoracic echo without evidence of a murmur on auscultation is referred to as “subclinical rheumatic heart disease.”[8]

Mitral regurgitation is the most common presentation of rheumatic heart disease in young people. However, rheumatic heart disease is the most common cause of mitral stenosis worldwide.[9] Common descriptions of the mitral valve on echocardiography are ‘dog-leg’ ‘elbow’ or ‘hockey-stick’ deformities, which all help describe the thickening and restricted motion of the anterior mitral valve leaflet.[8]

In 2012, the World Heart Federation released criteria for echocardiographic diagnosis of rheumatic heart disease. The criteria are dependent on age and are broken down into older or younger than 20 years old.

### **Echocardiographic Criteria for Individuals Aged Less Than or Equal to 20 Years Old**

#### **Definite Rheumatic Heart Disease (either A, B, C, or D)**

A: Pathological mitral regurgitation and at least two morphological features of rheumatic heart disease of the mitral valve

B: Mitral stenosis mean gradient greater than or equal to 4 mmHg

C: Pathological aortic regurgitation and at least two morphological features of rheumatic heart disease of the aortic valve

D: Borderline disease of both the aortic valve and mitral valve

#### **Borderline Rheumatic Heart Disease (either A, B, or C)**

A: At least two morphological features of rheumatic heart disease of the mitral valve without pathological mitral regurgitation or mitral stenosis

B: Pathological mitral regurgitation

C: Pathological aortic regurgitation

#### **Normal Echocardiographic Findings (all four)**

1. Mitral regurgitation not meeting all four Doppler echocardiographic criteria (physiological mitral regurgitation)
2. Aortic regurgitation not meeting all four Doppler echocardiographic criteria (physiological aortic regurgitation)
3. An isolated morphological feature of rheumatic heart disease of the mitral valve (for example, valvular thickening) without any associated pathological stenosis or regurgitation
4. The morphological features of rheumatic heart disease of the aortic valve (for example, valvular thickening) without any associated pathological stenosis or regurgitation

### **Echocardiographic Criteria for Individuals Aged Greater than 20 Years Old**

#### **Definite Rheumatic Heart Disease (any one of the four)**

1. Pathological mitral regurgitation and at least two morphological features of rheumatic heart disease of the

mitral valve

2. Mitral stenosis mean gradient greater than or equal to 4 mmHg
3. Pathological aortic regurgitation and at least two morphological features of rheumatic heart disease of the aortic valve, only in individuals aged less than 35 years
4. Pathological aortic regurgitation and at least two morphological features of rheumatic heart disease of the mitral valve

## **Morphological Features of Rheumatic Heart Disease**

### **Mitral Valve**

- Anterior mitral valve leaflet thickening greater than or equal to 3 mm (age-specific)
- Chordal thickening
- Restricted leaflet motion
- Excessive leaflet tip motion during systole

### **Aortic Valve**

- Irregular or focal thickening
- Coaptation defect
- Restricted leaflet motion
- Prolapse

### **Pathologic Mitral Regurgitation (All four Doppler criteria must be met)**

- Seen on two views
- On at least one view jet length greater than or equal to 2 cm
- Peak velocity of greater than or equal to 3 meters/second
- Pansystolic jet in at least one envelope

### **Pathologic Aortic Regurgitation (All four Doppler criteria must be met)**

- Seen on two views
- On at least one view jet length greater than or equal to 1 cm
- Peak velocity greater than or equal to 3 meters/second
- Pandiastolic jet in at least one envelope

## **Treatment / Management**

Management of rheumatic heart disease can be broken down into prevention and long-term management. Primary prevention of rheumatic heart disease centers on speedy recognition and treatment of group A streptococcal pharyngitis to prevent the development of acute rheumatic fever. Intramuscular benzathine penicillin G is the most widely used antibiotic to treat group A streptococcal pharyngitis.[10]

If a patient has a proven diagnosis of acute rheumatic fever, the goal of treatment is to suppress the inflammatory response to minimize the effects of inflammation on the heart and joints. According to the World Heart Federation, the only cost-effective approach to preventing the progression of rheumatic heart disease is secondary prophylaxis in the form of penicillin injections every 3 to 4 weeks to prevent recurrent group A streptococcal infection that causes recurrent episodes of acute rheumatic fever, which leads to progression of rheumatic heart disease.[8][10] The optimal duration of secondary prevention is unknown.[10]

Percutaneous mitral balloon valvuloplasty is the standard first-line therapy for cases of rheumatic mitral stenosis in the absence of regurgitation, arrhythmias, and left atrial thrombus.[9] Surgical intervention is commonly the mainstay of treatment in severe cases of valvular disease. Surgical intervention can either be valve replacement or repair.

If a patient develops heart failure due to valve disease, the patient should be placed on medical therapy as tolerated for heart failure including angiotensin-converting enzyme (ACE) inhibitors, diuretics, and beta-blockers.

At this time, there is no cure for rheumatic heart disease.

Clinicians should also have appropriate surveillance of patients diagnosed with rheumatic fever and rheumatic heart disease to allow for the initiation of appropriate therapy as early as possible.

## Differential Diagnosis

- Endocarditis
- Functional mitral regurgitation associated with viral illness, such as viral myocarditis
- Mitral valve prolapse due to degenerative myxomatous disease

## Prognosis

Rheumatic heart disease causes at least 200000 to 250000 premature deaths every year.[6] It is also the major cause of cardiovascular death in children and young adults in countries that have poor medical attention. If the valvular disease is not serially monitored, the patient may not present until severe heart failure is present resulting in a surgical contraindication.

## Complications

Rheumatic heart disease is generally latent or silent until cardiac complications develop in late adulthood. Infective endocarditis, an embolic event, heart failure, pulmonary hypertension, and atrial fibrillation from untreated severe valvular disease are the most common complications.[10]

## Deterrence and Patient Education

Rheumatic heart disease is highly prevalent in developing countries leading to the cause of the most cardiovascular morbidity and mortality in young people. Guidelines provided by the World Health Federation have outlined that the most cost-effective way to prevent rheumatic heart disease is with antibiotic prophylaxis after the diagnosis of rheumatic fever. Patients should be informed of their diagnosis of rheumatic fever as well as the complications associated with poor compliance with antibiotics and follow-up including valvular heart disease, heart failure, and arrhythmias.

## Enhancing Healthcare Team Outcomes

Patients with rheumatic heart disease are best managed by an interprofessional team. Primary care clinicians who have patients with a known diagnosis of rheumatic fever should maintain and stress the importance of strict follow-up with the patients. To prevent rheumatic heart disease, patients should undergo close monitoring with complete history

and physicals and transthoracic echocardiography. Patients should be referred to cardiology if they start developing a new murmur, signs, and symptoms of heart failure, arrhythmia, or evidence of valvular disease on transthoracic echocardiography. Surgery is often required when the mitral and/or aortic valve is severely damaged.

## Review Questions

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