

Rheumatic Fever

Authored by
mohammad looti

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1. Core Definition

Rheumatic fever (RF) is a severe, systemic, non-suppurative inflammatory disease that develops as a delayed sequela to a prior pharyngeal infection caused by Group A *Streptococcus* (GAS), specifically *Streptococcus pyogenes*. Although often misconstrued merely as a simple bacterial complication, RF is fundamentally an autoimmune condition wherein the body's immune system, mobilized to fight the streptococcal antigens, mistakenly begins to attack host tissues due to a phenomenon known as molecular mimicry. This catastrophic inflammatory response can target several organ systems throughout the body, but it exhibits a profound tropism for the heart, joints, skin, and central nervous system. The acute phase of RF typically manifests two to four weeks following an inadequately treated or undiagnosed streptococcal pharyngitis, commonly presenting with fever and migratory polyarthritis.

The initial insult, the strep throat, is common and generally treatable, yet it is the subsequent systemic reaction that defines the serious morbidity associated with RF. The disease is classified as a diffuse connective tissue disorder, capable of causing widespread inflammation (pancarditis) that can involve all layers of the heart--the endocardium, myocardium, and pericardium. Historically and currently, the most significant long-term consequence is chronic damage to the heart valves, resulting in Rheumatic Heart Disease (RHD). This distinction is critical: RF is the acute illness triggered by the infection, while RHD represents the permanent structural damage left behind, often necessitating complex surgical interventions later in life, highlighting why early diagnosis and aggressive prevention of the initiating GAS infection remain paramount goals in global public health strategy.

While the clinical presentation of RF can be highly variable, ranging from subclinical, mild joint pain to life-threatening heart failure, the underlying immunological mechanism remains consistent. The severity of the disease and the likelihood of developing RHD are strongly correlated with the intensity of the carditis experienced during the initial acute episode. Furthermore, RF is known to recur upon subsequent streptococcal infections, meaning that each new untreated infection carries the risk of compounding cardiac damage, thereby reinforcing the need for continuous prophylactic antibiotic regimens in affected individuals long after the initial diagnosis has been made.

2. Etiology and Pathogenesis

The definitive initiating factor for rheumatic fever is infection with Group A *Streptococcus*, yet only certain rheumatogenic strains of this bacteria, characterized primarily by specific M protein types, are capable of triggering the subsequent autoimmune cascade. The M protein, a filamentous

coiled-coil protein found on the surface of *S. pyogenes*, is highly variable and functions as a major virulence factor, enabling the bacteria to evade phagocytosis. In susceptible individuals, components of the bacterial cell wall, particularly specific epitopes on the M protein, share structural similarities with host proteins found in cardiac muscle (myosin), heart valve tissue (laminin and keratin), and neuronal cells (dopamine receptors). This sharing of epitopes is the essence of **molecular mimicry**, which provides the critical link between the bacterial infection and the sterile, inflammatory response characteristic of RF.

Following an infection, the host immune system generates antibodies (particularly IgG) against the streptococcal antigens, aiming to neutralize the pathogen. However, due to molecular mimicry, these cross-reactive antibodies mistakenly bind to self-antigens in the heart and other tissues. This binding initiates both humoral and cell-mediated immune responses against the host's own structures. In the heart, this results in the formation of Aschoff bodies--pathognomonic microscopic nodules of inflammatory cells (T-lymphocytes, macrophages, and giant cells) found within the myocardium. These localized areas of necrosis and inflammation are central to the structural and functional damage observed during acute carditis, leading directly to valve leaflet thickening, fusion, and eventual regurgitation or stenosis.

The development of RF is not solely dependent on the presence of a rheumatogenic strain; it is also heavily influenced by host factors, suggesting a significant genetic predisposition. While the exact genetic markers are complex, certain human leukocyte antigen (HLA) Class II alleles have been associated with increased susceptibility. Furthermore, environmental factors, such as socioeconomic status, overcrowding, and poor access to healthcare--all contributing to repeated and poorly managed streptococcal infections--play a pivotal role. It is the combination of the specific rheumatogenic bacterial strain, a genetically susceptible host, and the environmental context that ultimately dictates the progression from a simple strep throat to the severe, potentially fatal, systemic sequelae of rheumatic fever.

3. Key Characteristics and Clinical Manifestations

The diagnosis of acute rheumatic fever relies traditionally on the revised **Jones Criteria**, a set of clinical and laboratory guidelines that categorize manifestations into 'major' and 'minor' criteria, along with evidence of a preceding streptococcal infection. The presentation is highly variable, reflecting the multi-system involvement of the autoimmune process. The classic major manifestations include carditis, polyarthritis, chorea, erythema marginatum, and subcutaneous nodules. Among these, carditis and arthritis are the most common initial complaints, requiring careful clinical differentiation from other systemic inflammatory diseases.

Polyarthritis, often the most common symptom, typically presents as a migratory, transient inflammation affecting large joints (knees, ankles, elbows, wrists). The migratory nature means the

inflammation subsides rapidly in one joint only to appear dramatically in another within hours or days. Crucially, rheumatic arthritis is almost always exquisitely responsive to nonsteroidal anti-inflammatory drugs (NSAIDs) or salicylates (aspirin) and leaves no permanent joint damage, differentiating it from conditions like rheumatoid arthritis. **Carditis**, however, is the most serious manifestation, occurring in approximately 50-70% of initial RF episodes. It can manifest as a new heart murmur, indicating valvular insufficiency (especially mitral regurgitation), pericardial friction rubs, or signs of congestive heart failure. The long-term prognosis hinges entirely on the presence and severity of this carditis.

Other less frequent but highly specific major manifestations include **Sydenham's chorea**, a neurological disorder characterized by purposeless, involuntary, jerky movements, often affecting the face and limbs, which can appear months after the initial infection and sometimes without other inflammatory signs. **Erythema marginatum** is a rare, non-pruritic rash consisting of pink rings with pale centers, primarily appearing on the trunk and proximal limbs. Finally, **subcutaneous nodules** are firm, painless lumps located over bony prominences, typically the elbows or knuckles, indicating severe disease activity, although they are now exceedingly rare in clinical practice. The presence of two major criteria, or one major and two minor criteria (such as fever, arthralgia, elevated acute phase reactants like C-reactive protein, and prolonged P-R interval on ECG), coupled with definitive evidence of a recent GAS infection, is required for a conclusive diagnosis.

4. Epidemiology and Historical Context

The global epidemiological landscape of rheumatic fever presents a stark duality. In industrialized, developed nations, such as the United States, Canada, and Western Europe, the incidence of acute RF has dramatically declined since the mid-20th century, largely attributed to widespread availability and utilization of antibiotics (penicillin) for treating streptococcal pharyngitis, improved hygiene, and enhanced socioeconomic conditions. In these areas, RF is now considered a rare disease, often only seen in localized outbreaks or among specific underserved populations where access to primary care is limited. However, despite this progress in the West, RF remains a devastating and persistent public health crisis in many low- and middle-income countries, particularly in sub-Saharan Africa, the Middle East, Central Asia, and parts of the Pacific region.

The sheer burden of RF in the developing world is staggering. It is estimated that Rheumatic Heart Disease, the chronic consequence of RF, affects over 33 million people globally and results in approximately 300,000 deaths annually, disproportionately impacting children and young adults under 25. The persistence of RF in these regions is fundamentally linked to socio-environmental factors that facilitate the transmission of GAS, including poverty, overcrowded living conditions, and inadequate sanitation. These conditions promote frequent upper respiratory tract infections and make timely antibiotic treatment impractical or inaccessible, leading to a high prevalence of untreated or inadequately treated streptococcal infections, which serves as the continuous

reservoir for RF development.

Historically, prior to the antibiotic era, RF was a common and feared disease worldwide, often referred to as the "disease that licks the joints but bites the heart." Its decline in developed nations stands as one of the major triumphs of 20th-century medicine and public health policy. However, the persistent, high prevalence rates in endemic regions underscore a critical global health inequity. Current international efforts, spearheaded by organizations like the World Health Organization ([WHO](#)), are focused on strengthening primary prevention programs--ensuring widespread access to penicillin for sore throats--and secondary prevention--maintaining antibiotic prophylaxis for those already diagnosed with RF to prevent recurrence and further cardiac damage.

5. Complications: Rheumatic Heart Disease

The most severe and life-altering complication of rheumatic fever is **Rheumatic Heart Disease (RHD)**, which results from permanent, irreversible damage to the heart valves following acute carditis. While the initial inflammation in the joints and skin is usually self-limiting and resolves without residual effects, the inflammation affecting the endocardium leads to progressive fibrosis, calcification, and destruction of the valvular tissue. The mitral valve is by far the most commonly affected (around 60-70% of cases), often leading to mitral regurgitation (leakage) during the acute phase, and later progressing to mitral stenosis (narrowing) years or decades later, which restricts blood flow and increases pressure in the left atrium.

A critical aspect of RHD pathogenesis is its cumulative nature. Each subsequent, untreated GAS infection in an individual who has previously experienced RF carries a high risk of triggering a recurrent episode of carditis. These recurrent episodes layer inflammatory damage onto already scarred valves, accelerating the transition from mild valvular dysfunction to severe, hemodynamically significant stenosis or regurgitation. This progressive pathology ultimately leads to chronic conditions such as pulmonary hypertension, atrial fibrillation, and severe congestive heart failure, necessitating complex and costly interventions like lifelong anticoagulation, percutaneous valvuloplasty, or outright surgical valve replacement.

The long latency period between the initial RF episode in childhood or adolescence and the clinical presentation of severe, symptomatic RHD in early to mid-adulthood creates significant diagnostic challenges in endemic areas. Patients may present with advanced heart failure having long forgotten the childhood illness that initiated the damage. Given that surgical intervention for RHD is expensive and requires highly specialized medical infrastructure, the high prevalence of RHD in resource-poor settings generates an immense strain on fragile healthcare systems, perpetuating a cycle of illness and economic hardship that extends far beyond the immediate medical consequences of the disease.

6. Treatment and Prevention

The management of rheumatic fever is typically bifurcated into two phases: treating the acute episode and providing long-term prevention against recurrence. Treatment for acute RF focuses primarily on suppressing the systemic inflammation and providing supportive care. Anti-inflammatory agents are essential; high-dose aspirin or other NSAIDs are the standard treatment for arthritis, providing rapid symptom relief. For patients with moderate to severe carditis, corticosteroids (e.g., prednisone) are often utilized to reduce severe myocardial inflammation and potentially minimize long-term cardiac damage, although their definitive role in preventing RHD remains a subject of ongoing clinical debate. In all cases, even if the primary pharyngeal infection has apparently resolved, a course of antibiotics (usually penicillin) is administered immediately upon diagnosis of RF to eradicate any residual GAS organisms and prevent the potential shedding of bacteria that could sustain the immune reaction.

The cornerstone of controlling the RF burden globally lies in **prevention**, categorized into primary and secondary strategies. Primary prevention involves the timely and appropriate treatment of all cases of streptococcal pharyngitis with a full course of antibiotics (typically a single injection of Benzathine penicillin G or oral penicillin V) within nine days of symptom onset. This strategy prevents the initial episode of RF entirely. Unfortunately, adherence to full antibiotic courses and accurate diagnosis of strep throat remain barriers in many communities.

Secondary prevention is arguably the most critical clinical intervention for individuals already diagnosed with RF, particularly those with established RHD. This involves the continuous administration of prophylactic antibiotics--usually monthly intramuscular injections of Benzathine penicillin G--to prevent recurrent streptococcal infections, which, as previously noted, trigger exacerbations of carditis and hasten the progression of RHD. The duration of this prophylaxis is extensive, ranging from five years or until age 21 (for RF without carditis) to lifetime prophylaxis for those with severe RHD or a history of recurrent RF. Adherence to this arduous long-term regimen, particularly in resource-limited settings, presents substantial logistical and socioeconomic challenges, which public health programs are continuously working to overcome through patient education and robust supply chains.

7. Further Reading

[Rheumatic Fever - Wikipedia](#)

[Rheumatic Fever - Centers for Disease Control and Prevention \(CDC\)](#)

[Rheumatic fever and rheumatic heart disease - World Health Organization \(WHO\)](#)

[Acute Rheumatic Fever - StatPearls Publishing \(NCBI\)](#)