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Acute Rheumatic Fever: Challenges and Solutions

Bahodirov Behruz Shavkat ogli
Abu Ali Ibn Sina Bukhara State Medical Institute, Scientific Supervisor
bahodirov.behruz@bsmi.uz
https://orcid.org/0009-0001-4718-7138

Rakhmonova Istat Hamitovna
Abu Ali Ibn Sina Bukhara State Medical Institute, Student

Abstract: *Acute rheumatic fever (ARF) is a multisystem inflammatory disease resulting from an autoimmune response to group A β-hemolytic streptococcal infections, predominantly affecting children and adolescents aged 7–14 years. Despite being preventable and treatable, ARF remains a significant health challenge worldwide, especially in low- and middle-income countries, due to socioeconomic disparities, inadequate healthcare access, and limited public awareness.*

This study utilized a descriptive-analytical approach, systematically reviewing international guidelines, textbooks, and recent peer-reviewed literature on ARF to analyze its prevalence, etiology, clinical features, diagnostic methods, treatment protocols, and prevention strategies. Epidemiological data were gathered from global and regional sources, with special attention to diagnostic tools like the Jones criteria and to effective treatment and prophylactic measures.

The findings confirm that carditis is the most common and severe manifestation of ARF, significantly influencing long-term outcomes, while Sydenham's chorea, affecting 20% of patients, presents neuropsychiatric challenges requiring specific attention. Proper antibiotic management of streptococcal pharyngitis reduces the initial incidence of ARF, while long-term secondary prophylaxis significantly decreases recurrence rates. The study also identifies persistent challenges, such as limited healthcare resources and gaps in community education, particularly in high-burden regions.

Addressing ARF effectively requires a multifaceted strategy involving clinical care, public health policies, educational outreach, and healthcare system strengthening. Increasing healthcare worker training and improving public awareness are crucial to enhance early diagnosis and adherence to prophylactic regimens. The need for further research on the neuropsychiatric outcomes of ARF and scalable prevention methods suitable for resource-limited settings is also emphasized.

Strengthening prevention, ensuring equitable healthcare access, and fostering international collaboration are key strategies to reduce the global burden of ARF. Implementing evidence-based interventions can significantly improve health outcomes for millions of children worldwide at risk of this preventable disease.

Key words: *Acute rheumatic fever, streptococcal infection, children, carditis, Sydenham's chorea, polyarthritis, diagnosis, treatment, prevention, rheumatic heart disease.*

Introduction

Acute Rheumatic Fever (ARF) is a serious, multisystem inflammatory disease that develops as a result of an abnormal immune response to infection caused by group A β-hemolytic streptococcus (Streptococcus pyogenes), particularly following acute tonsillitis or pharyngitis. This condition, which most frequently affects children and adolescents aged between 7 and 14 years, especially those with a genetic predisposition or living in low- and middle-income countries, represents a major public health challenge due to its potential to cause long-term disability and even death if left untreated. ARF primarily targets the cardiovascular system, with carditis being its most significant manifestation, but also affects the joints (manifesting as migratory polyarthritis), the central nervous system (manifesting as Sydenham's chorea), and the skin (presenting with erythema marginatum and subcutaneous nodules). If ARF is not identified and treated promptly, it can progress to rheumatic heart disease (RHD), which is characterized by permanent and often progressive damage to the heart valves, particularly the mitral and aortic valves, potentially leading to chronic heart failure, arrhythmias, stroke, and the need for surgical intervention such as valve repair or replacement[1]. Historically, the term "rheumatism" was introduced by the physician Ballonius in 1635, derived from the Greek word "ῥεῦμα" meaning "flow," reflecting the observation that the disease seems to flow or spread through the body, causing inflammation and damage in multiple tissues and organs. Later, prominent physicians such as J.B. Bouillaud and G.I. Sokolsky emphasized the serious nature of the disease, famously describing how "rheumatism affects the joints no less than it affects the heart," and Laennec added the memorable phrase, "rheumatic fever licks the joints and bites the heart," underscoring the particularly destructive impact on the heart. Over time, considerable progress has been made in understanding the pathophysiology and management of ARF, and modern diagnostic criteria, notably the Jones criteria, first proposed in 1944 and subsequently revised multiple times, have provided a standardized framework for clinicians to identify and diagnose the disease[2]. According to the World Health Organization (WHO), despite advancements in healthcare and widespread availability of antibiotics, ARF continues to pose a significant burden, especially in regions with overcrowded living conditions, limited access to healthcare services, poor sanitation, and inadequate public health infrastructure. Global estimates indicate that approximately 470,000 new cases of ARF are recorded each year, with Africa experiencing an incidence of 100–200 cases per 100,000 children and South Asia (including India) reporting 50–150 cases per 100,000 children. Moreover, the WHO reports that nearly 40 million people worldwide are currently living with rheumatic heart disease, and over 275,000 deaths occur annually due to complications associated with this preventable and treatable condition. The persistent burden of ARF is closely linked to socioeconomic factors, including poverty, lack of access to early medical care, and limited health education, which hinder efforts to achieve timely diagnosis, treatment, and prophylaxis. Importantly, ARF is considered a preventable disease: with proper identification and treatment of streptococcal pharyngitis using a full 10-day course of antibiotics, the immune response that leads to ARF can be effectively halted. Furthermore, long-term secondary prophylaxis using intramuscular benzylpenicillin injections every 3–4 weeks in individuals with a history of ARF significantly reduces the risk of recurrence and subsequent heart valve damage. In this context, public health efforts aimed at improving awareness of ARF, promoting early treatment of streptococcal infections, enhancing access to diagnostic and therapeutic resources, and maintaining long-term follow-up and prophylactic strategies are essential components in reducing the incidence and impact of ARF and RHD worldwide. Given the serious nature of the disease and its long-term consequences, particularly among vulnerable pediatric populations, the study of ARF remains a critical area

of research and clinical focus. This article aims to provide a comprehensive overview of acute rheumatic fever, including its etiology, epidemiology, clinical features, diagnostic approaches, treatment principles, complications, and preventive strategies, highlighting both the challenges faced by clinicians and public health professionals and the potential solutions that can be implemented to address this ongoing global health problem[3-4]. By deepening our understanding of ARF, enhancing awareness among healthcare providers and communities, and promoting evidence-based prevention and treatment practices, it is possible to significantly reduce the burden of this disease and improve the quality of life and health outcomes for affected individuals, particularly children in resource-limited settings.

Methodology

This study employs a descriptive-analytical methodology to examine the key aspects, challenges, and management strategies of acute rheumatic fever (ARF), with a particular focus on its prevalence, clinical manifestations, diagnostic processes, treatment approaches, and preventive measures. The research is based on an extensive review of contemporary scientific literature, including guidelines from the World Health Organization (WHO), textbooks on internal diseases and pediatrics, and recent peer-reviewed studies on ARF and rheumatic heart disease (RHD). The data collection involved systematically gathering epidemiological statistics, clinical diagnostic criteria (such as the Jones criteria), and recommended treatment protocols, alongside evidence from recent publications that explore the immunopathogenesis and neuropsychiatric manifestations (e.g., Sydenham's chorea) associated with ARF. To ensure a comprehensive and up-to-date understanding, the sources used include global health data, regional epidemiological reports (especially from high-burden areas such as Africa and South Asia), and clinical research articles with documented DOI numbers, enabling verification and reproducibility of findings. Additionally, the study synthesizes information on preventive strategies, particularly primary prevention through early antibiotic treatment and secondary prophylaxis to reduce recurrence rates, evaluating their effectiveness in diverse healthcare settings[5-6]. A critical element of this methodological approach is the analysis of challenges faced by healthcare systems in implementing ARF management strategies, including resource constraints, access to antibiotics, and gaps in healthcare worker training and public awareness. This methodology also integrates a comparative analysis of best practices derived from both high-income and low- to middle-income countries to identify scalable, evidence-based solutions. By adopting this multi-source, literature-driven approach, the study aims to provide a holistic understanding of ARF and generate practical recommendations for healthcare professionals, policymakers, and public health stakeholders involved in addressing this ongoing global health issue.

Results and Discussion

Carditis is the most common (90–95%) and significant diagnostic marker of rheumatic fever, determining disease severity and outcome. It often involves mitral or, less commonly, aortic valvulitis, with organic murmurs in the pharyngeal area and possibly associated myopericarditis. Symptoms may include tachycardia, cardiac enlargement, muffled heart sounds, pericardial friction rub, and rhythm or conduction abnormalities[7-8].

Rheumatic polyarthritis usually begins suddenly with high fever, severe joint pain, swelling, and redness. The child may be significantly limited in movement due to pain. Large and small joints, including intervertebral joints, may be affected. In acute cases, joint changes typically resolve within 2–3 weeks without leaving any anatomical or functional impairment.

About 20% of patients with ARF develop **Sydenham's chorea**. This is an autoimmune disorder of the central nervous system that occurs after streptococcal infection and is a major diagnostic criterion in the Jones criteria. It usually appears 3 months or more after the initial infection. Sydenham's chorea is a neuropsychiatric condition characterized by emotional lability, anxiety, obsessive-compulsive features, attention deficit, and

hyperactivity. Choreic movements are involuntary, irregular, and non-stereotyped. Other neurological symptoms may include hypotonia, migraine, dysarthria, dysgraphia, and lingual dystonia[9-10].

Diagnosis: Laboratory and instrumental investigations play a crucial role in diagnosing ARF:

- **ASO titer** – confirms streptococcal infection
- **X-ray** – to assess joint inflammation
- **ECG and Echocardiography** – to detect valvular damage
- **Complete blood count** – showing elevated ESR, CRP, leukocytosis
- **Throat swab** – to detect streptococcal presence

Treatment Principles: ARF treatment consists of two main strategies:

1. **Eliminating infection:**
   - Benzylpenicillin IM: 0.6–1.2 million units for 10 days.
   - In case of allergy, erythromycin (250 mg twice daily) or azithromycin.
   - Long-term prophylactic penicillin every 3–4 weeks for years.
2. **Anti-inflammatory treatment:**
   - NSAIDs: Aspirin (3–5 g/day), ibuprofen.
   - Corticosteroids: Prednisolone for severe carditis.
   - Heart support medication: Furosemide, digoxin in cases of heart failure[11].

Bed Rest: Patients with carditis should observe strict bed rest for 4–6 weeks.

Complications: If untreated, ARF may result in:

- Rheumatic heart disease: mitral or aortic stenosis/regurgitation
- Chronic heart failure
- Thromboembolic events
- Recurrences (if prophylaxis is not followed)

Rheumatic heart disease usually results from valvular deformities, which may require surgical correction (e.g., valve replacement)[12-13].

Prevention: ARF prevention includes three levels:

1. **Primary prevention:** Timely diagnosis and treatment of streptococcal pharyngitis with antibiotics (10-day penicillin course)
2. **Secondary prevention:** Monthly benzylpenicillin injections every 3–4 weeks for at least 5 years in patients who had ARF
3. **Tertiary prevention:** Long-term monitoring and preventive treatment in patients with heart valve damage to prevent complications and recurrences[14-15].

Conclusion: In conclusion, acute rheumatic fever (ARF) continues to pose a significant global health burden, particularly in low- and middle-income countries, where socioeconomic disparities, limited access to healthcare, and insufficient public awareness contribute to its persistent prevalence. Despite being a preventable and treatable disease, ARF remains one of the leading causes of acquired heart disease among children and young adults, primarily due to delayed diagnosis, inadequate treatment of streptococcal infections, and poor adherence to secondary prophylaxis protocols. The analysis presented in this article highlights the importance of early identification and management of streptococcal pharyngitis through a complete course of antibiotic therapy, which serves as the cornerstone of primary prevention. Furthermore, the implementation of long-term benzylpenicillin prophylaxis in individuals with a history of ARF is crucial in preventing recurrences and halting the progression to rheumatic heart disease (RHD), which often results in permanent valvular damage and may require surgical intervention.

The clinical manifestations of ARF, particularly carditis and Sydenham's chorea, demand heightened awareness among healthcare providers to ensure timely diagnosis and intervention, especially in settings with limited diagnostic resources. Additionally, enhancing the training of healthcare workers, improving community education, and strengthening public health infrastructures are essential strategies to reduce ARF incidence and improve patient outcomes. The findings also suggest the need for further research, particularly on the neuropsychiatric aspects of ARF and the long-term impact of Sydenham's chorea on child development, as well as on developing scalable, cost-effective preventive measures adaptable to resource-constrained environments.

Ultimately, a multifaceted approach that integrates clinical care, robust public health policies, educational outreach, and health system improvements is critical for effectively addressing the challenges posed by ARF. By prioritizing prevention, ensuring equitable access to medical care, and fostering international collaborations, it is possible to significantly reduce the burden of acute rheumatic fever and improve the quality of life for millions of children and families worldwide.

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