

## HEALTH PROMOTION AND PREVENTIVE CARDIOLOGY

### CASE REPORT: CLINICAL CASE

# Rheumatic Fever Unveiled in a 13-Year-Old Misdiagnosed With Appendicitis



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### ABSTRACT

Rheumatic fever (RF) remains a significant global health concern, particularly in regions with limited access to health care. Despite its association with group A streptococcal infections, diagnosing RF can be challenging because of its diverse clinical presentations, including cardiac manifestations such as heart block. We present the case of a 13-year-old boy initially misdiagnosed with appendicitis whose subsequent evaluation revealed acute RF (ARF) with associated cardiac complications, including Mobitz type II heart block. Comprehensive cardiac investigations confirmed the diagnosis, highlighting the importance of considering ARF in patients presenting with cardiac abnormalities. Early recognition and management of ARF are crucial, particularly in cases involving cardiac manifestations such as heart block. Favorable outcomes were observed with appropriate medical therapy, emphasizing the effectiveness of multidisciplinary management in preventing disease progression. (JACC Case Rep. 2024;29:102632) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

**R**heumatic fever (RF) results from an inflammatory response initiated by group A streptococcal pharyngitis infections, potentially

leading to rheumatic heart disease, characterized by cardiac valve damage.<sup>1</sup> Despite advancements in health care, RF remains prevalent globally, particularly in developing regions and specific demographic groups like the indigenous Māori and Pacific Islanders in New Zealand.<sup>2-4</sup>

Diagnosis of acute RF relies on clinical criteria involving major and minor symptoms.<sup>5</sup> Although RF is often perceived as a delayed autoimmune response to group A streptococcal pharyngitis, its precise pathogenesis remains elusive. The condition presents as a systemic inflammatory process affecting various organs, including the heart, joints, central nervous system, subcutaneous tissue, and skin. Although noncardiac manifestations typically resolve without lasting effects, cardiac involvement (carditis) can lead to disturbances in the cardiac conduction system,

### TAKE-HOME MESSAGES

- This case highlights the critical need for early recognition of rheumatic fever, particularly when cardiac manifestations such as heart block are present.
- Multidisciplinary management is imperative in preventing progressive cardiac complications in rheumatic fever.
- Abdominal pain mimicking appendicitis can be an atypical presentation of rheumatic fever, necessitating careful differential diagnosis to avoid misdiagnosis.

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**ABBREVIATIONS  
AND ACRONYMS**

**AV** = atrioventricular  
**ECG** = electrocardiogram  
**RF** = rheumatic fever

ranging from first-degree atrioventricular (AV) block to second-degree AV block, with rare instances of complete heart block.<sup>6</sup>

**CASE PRESENTATION**

A 13-year-old boy was referred from a peripheral hospital for appendectomy because of an unavailability of beds. Initial investigations at the peripheral hospital revealed a 5-mm appendix without free fluid on ultrasound. However, irregularities in pulse and electrocardiographic findings prompted surgery cancellation and referral to King Fahad Central Hospital in Jazan. The patient had a history of palpitations with exercise for 6 months but no previous cardiac disease. On examination, he presented with abdominal pain in the right iliac fossa, associated with vomiting, loss of appetite, burning micturition, and newly developed diarrhea. Despite stable vital signs, the patient exhibited bradycardia (45-52 beats/min) without murmurs detected.

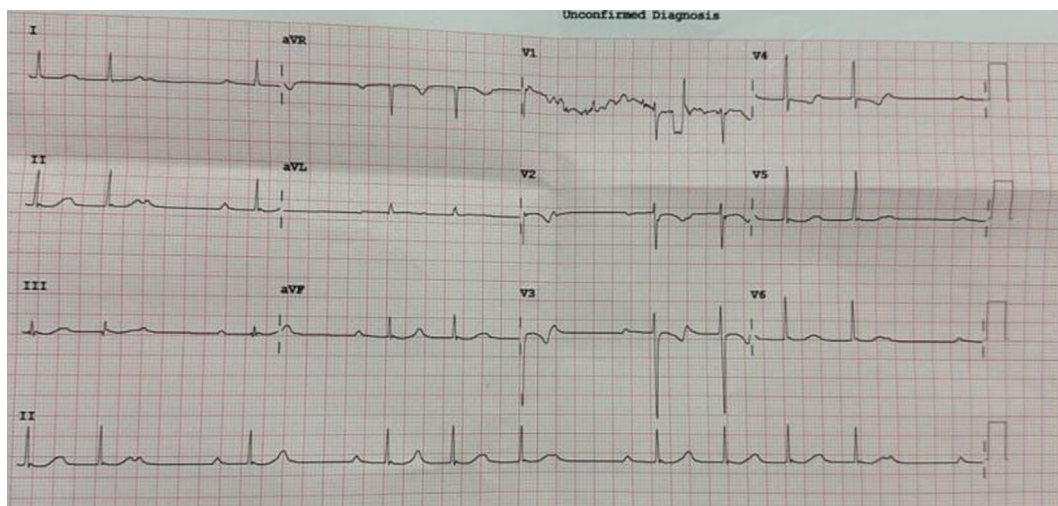
The abdominal examination revealed tenderness in the right iliac fossa but no rebound tenderness or rigidity. On assessment, suspected acute appendicitis along with bradycardia and second-degree Mobitz type II heart block were noted and confirmed by electrocardiogram (ECG) (Figure 1). Cardiac investigations, including antistreptolysin O titer, anti-Dnase B, creatine kinase-myocardial band, erythrocyte sedimentation rate, and C-reactive protein, were conducted, revealing significant results. The erythrocyte sedimentation rate was 70 mm/h,

C-reactive protein was 20.2 mg/dL, and anti-streptolysin O was 1,450 IU/mL; blood culture and urine culture showed no growth.

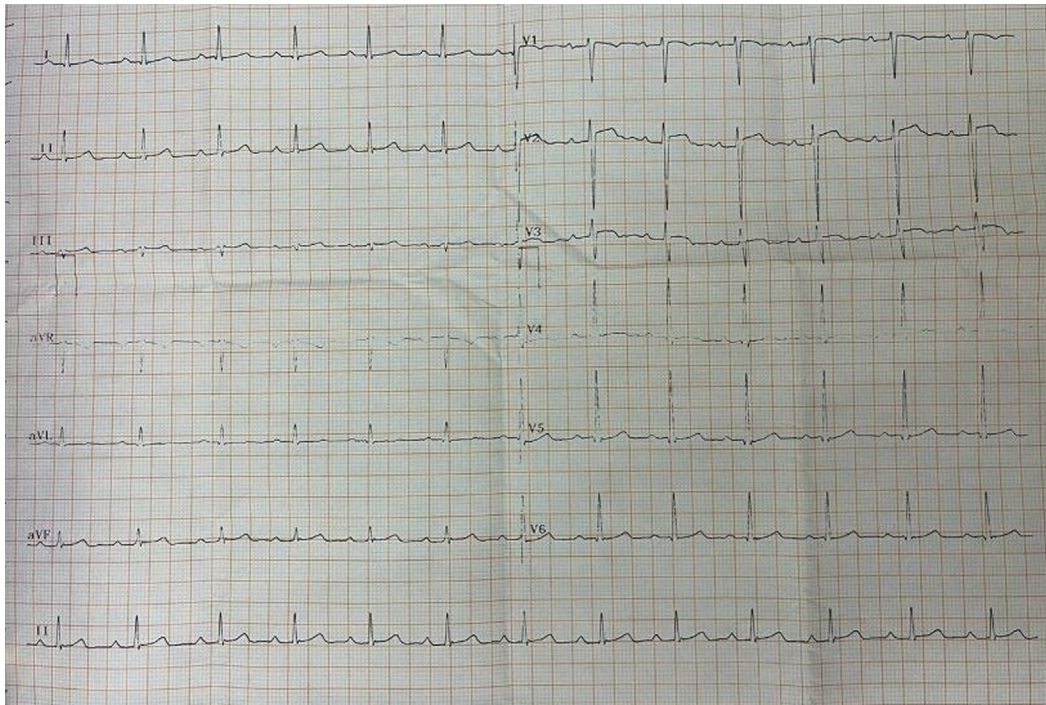
Cardiology findings indicated mild to moderate aortic regurgitation, mild mitral regurgitation, trivial tricuspid regurgitation, and a mild decrease in left ventricular function, with Mobitz type II heart block observed on ECG. Complete blood count results showed a white blood cell count of  $10.28 \times 10^3/\mu\text{L}$  (with neutrophil 68.5%, lymphocyte 21.8%, and monocyte 9.2%), red blood cell count of  $4.74 \times 10^6/\mu\text{L}$ , hemoglobin of 10.8 g/dL, and platelet count of  $318 \times 10^3/\mu\text{L}$ . The basic metabolic profile was within normal range, and urine culture showed no growth. These laboratory findings provided valuable insight into the inflammatory process and aided in confirming the diagnosis of acute RF with associated cardiac complications.

**TREATMENT**

On confirmation of RF with associated cardiac complications, the patient received intravenous antibiotics and was initiated on a tapering regimen of prednisolone. Benzathine benzylpenicillin injections were administered, and oral medications including omeprazole were prescribed. In view of 1 major (carditis) plus 2 minor components (fever, elevated C-reactive protein) of the revised Jones criteria, along with evidence of previous streptococcal infection in the form of high antistreptolysin O titers, the diagnosis of acute RF was confirmed. The treatment

**FIGURE 1** Electrocardiogram Demonstrating Mobitz Type II Heart Block

**FIGURE 2** Electrocardiogram Demonstrating Resolution of Bradycardia and Normalization of Cardiac Rhythm



regimen aimed to address the underlying inflammatory process and cardiac abnormalities while also managing symptoms and preventing recurrence of RF.

### FOLLOW-UP

After discharge, the patient continued the prescribed medication regimen and attended regular follow-up appointments. Repeat echocardiography and cardiac monitoring were performed to assess treatment response and cardiac function. The prednisolone dosage was gradually tapered over 2 months, in conjunction with benzathine benzylpenicillin injections administered every 3 weeks. The patient showed improvement in symptoms and cardiac function during follow-up visits, with resolution of bradycardia and normalization of cardiac rhythm observed on subsequent ECGs (Figure 2). Long-term monitoring and management were planned to prevent disease progression and ensure optimal cardiac health.

### DISCUSSION

Abdominal pain is a nonspecific and relatively uncommon symptom of acute RF, occurring in <5% of

patients. When it does appear, it typically precedes other rheumatic signs by a few hours or days. Generally mild and short-lived, it can occasionally be severe enough to mimic appendicitis.<sup>7,8</sup>

Distinguishing rheumatic abdominal pain from early acute appendicitis can be challenging, especially when abdominal symptoms precede other rheumatic manifestations. In our patient, abdominal ultrasonography was instrumental in establishing the correct diagnosis. Kula et al<sup>9</sup> reported a similar case initially diagnosed as acute appendicitis; the patient developed arthritis and carditis a few days post-appendectomy, with pathologic examination confirming true inflammation of the appendix. In our reported case, we did not proceed to appendectomy because the patient developed electrocardiographic abnormalities, which could be related to acute RF. Given the potential risks, it is often preferable to remove a normal appendix than to risk rupture and generalized peritonitis.<sup>10</sup>

Our case also presented an unusual finding of a Mobitz type II AV block on ECG. The diagnosis of acute RF was supported by a history of low-grade fever, arthritis, clinical and echocardiographic evidence of carditis, and laboratory confirmation of streptococcal infection. Treatment with steroids led

to the patient regaining normal sinus rhythm within 2 days, similar to the outcomes reported by Kula et al.<sup>9</sup> Although first-degree heart block is a common electrocardiographic manifestation of acute RF and included in the revised Jones criteria, more advanced blocks are rare.<sup>11</sup>

The initial presentation of appendicitis-like symptoms in our 13-year-old patient posed a diagnostic challenge because of overlapping clinical features. However, the identification of bradycardia and Mobitz type II heart block on ECG raised suspicion of cardiac involvement, ultimately leading to the diagnosis of acute RF with associated cardiac complications. RF, commonly caused by group A streptococcus infections, is known for its cardiac implications.<sup>12</sup> Although first-degree AV block predominates in acute RF, severe AV conduction abnormalities like Mobitz type I and complete AV block have also been reported.<sup>6,13</sup> Incidence rates for first-degree AV block during acute RF episodes range between 34.2% and 72.3%.<sup>14,15</sup> Additionally, advanced AV blocks, junctional rhythm, premature atrial contractions, ventricular extrasystoles, and ventricular/supraventricular tachycardias have been observed in acute RF patients.<sup>10,16</sup>

Cardiac manifestations in acute RF range from first-degree AV block to complete heart block, with varying incidence rates documented in the literature.<sup>6,13</sup> The presence of Mobitz type II heart block in our patient, alongside significant cardiac valve abnormalities identified through echocardiography, highlights the severity of acute RF-associated cardiac complications. Moreover, the occurrence of complete heart block during a recurrent episode of RF in a patient with established rheumatic heart disease emphasizes the potential for disease progression and the need for vigilant monitoring.<sup>10,17</sup> Zalstein et al<sup>10</sup> reported first-degree AV block in 72.3% of acute RF patients, Mobitz type I AV block in 1.5%, and complete AV block in 4.6%. Agnew et al<sup>16</sup> documented various transient AV conduction abnormalities in 8.5% of patients, with second- and third-degree AV block rates at 2.5%. Rare cases have been reported both Mobitz type II and third-degree AV block in the same acute RF episode.<sup>18,19</sup>

Electrocardiographic abnormalities, particularly AV block, are frequently documented in acute RF cases, with various degrees of conduction disturbances reported in the literature.<sup>13</sup> The manifestation of Mobitz type II heart block in our patient underscores the importance of meticulous cardiac monitoring in suspected cases of acute RF,

particularly in the context of recurrent symptoms or established rheumatic heart disease. Although the exact pathogenesis of AV block in acute RF remains unclear, increased vagal tone and immunologic effects on the AV node have been proposed as potential causes.<sup>17</sup> Conduction abnormalities associated with acute RF are typically self-limited and often improve within several weeks after initiation of nonsteroidal anti-inflammatory therapy.<sup>20</sup> Corticosteroids have also shown promise in treating advanced AV block due to acute RF.<sup>21</sup> Nonetheless, cases requiring permanent pacemaker implantation have been reported, particularly in instances of syncope and hemodynamic instability.<sup>22</sup>

The incidence of acute RF-related AV block underscores the importance of considering acute RF in patients presenting with various degrees of AV block. Continuous rhythm monitoring, such as 24-hour Holter recordings, has proven invaluable in detecting rhythm abnormalities in acute RF patients.<sup>23,24</sup>

Treatment strategies for acute RF aim to mitigate the underlying inflammatory process and prevent RF recurrence, typically involving a combination of antibiotics, anti-inflammatory agents, and cardiac management.<sup>20</sup> In our case, the patient received intravenous antibiotics, corticosteroids, and benzathine benzylpenicillin injections, with favorable outcomes observed during subsequent follow-up assessments.

## CONCLUSIONS

This case underscores the critical need for early recognition of RF, especially when cardiac manifestations such as heart block are present. Prompt diagnosis and multidisciplinary management are imperative to prevent progressive cardiac complications. Furthermore, further research into the pathogenesis of heart block in acute RF and the efficacy of novel treatment modalities is warranted to enhance our understanding and optimize patient care.

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**KEY WORDS** acute rheumatic fever, cardiac complications, diagnosis, heart block, rheumatic fever