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Severe Rheumatic Heart Disease Requiring Mechanical Valve Placement in a Special Operations Forces Soldier

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ABSTRACT

Rheumatic heart disease (RHD) has become exceedingly rare in the United States, but a recent resurgence may place military Servicemembers at increased risk for this diagnosis. Our case describes a 29-year-old U.S.-born Special Operations Forces (SOF) Soldier who presented for recurrent exertional near-syncope and progressive exercise intolerance with subsequent workup remarkable for RHD. Initial electrocardiogram was notable for benign early repolarization and intraventricular conduction delay. Cardiology evaluation with transthoracic and transesophageal echocardiograms revealed severe mitral regurgitation and rheumatic appearing mitral valve leaflets. The patient underwent a successful mechanical mitral valve replacement, requiring lifelong anticoagulation with warfarin. Depending on severity of valvular disease, treatment modalities range from conservative medical therapies to invasive and minimally invasive surgical intervention. This case demonstrates the importance for SOF medics and providers to remain vigilant of this resurging disease process. Additionally, it emphasizes the necessity for a high level of clinical suspicion in those with exertional complaints and decreased exercise tolerance to ensure timely diagnosis and treatment of rare but potentially life-threatening conditions.

Keywords: rheumatic heart disease; exertional syncope; palpitations; mechanical valve; special operations

Introduction

Rheumatic heart disease (RHD) is a very rare sequela of strep pharyngitis and acute rheumatic fever (ARF) that can lead to valvular damage and potentially the need for cardiac surgery. Though exceedingly rare in the United States, it remains one of the most common acquired heart diseases of children and adolescents globally with a prevalence of over 40 million cases, accounting for over 300,000 deaths annually worldwide. In developed countries, including the U.S., ARF cases remain low with an incidence of <2 cases per 100,000 people and RHD-related deaths of 3.1 cases per 100,000 people. However, age-adjusted mortality rates have increased in the U.S. over the past 5 years with a suspected surge in RHD-related health

complications.3 Despite resolution of streptococcal pharyngitis and ARF, a chronic systemic immune response damages valves, leading most commonly to mitral stenosis (MS) or mitral regurgitation (MR).4 Individuals affected by RHD typically present between the ages of 20-30 years after an initial episode of ARF with symptoms of orthopnea, palpitations, exertional dyspnea, syncope, near-syncope, or decreased exercise tolerance.5 High-intensity exercise can accelerate progression of valvular disease; those in physically demanding jobs, such as military service or Special Operations, are at increased risk of exertional syncope, provoking underlying arrhythmias and potentially sudden cardiac death.6 While RHD is a rare diagnosis in the U.S., it carries significant morbidity and operational implications if undiagnosed and untreated. Therefore, military Servicemembers and those within Special Operations Forces (SOF) who present with palpitations, syncope, or decreased exercise tolerance should undergo a rigorous and focused evaluation to include these insidious cardiac etiologies.

Case Presentation

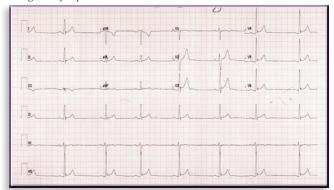
A 29-year-old active-duty male SOF Soldier presented to his Battalion Aid Station (BAS) for recurrent, intermittent near-syncope during exertion, increasing in frequency. Three years prior, the patient first noted exertional near syncope lasting 1-2 minutes, with spontaneous resolution and no loss of consciousness. He sought care at the BAS, though no further workup was pursued at that time. He returned to the BAS for more frequent episodes of near-syncope, which he described as tunnel vision, dizziness, seeing stars, and feel lightheaded, along with worsening exercise tolerance. Previously, the patient could run seven to eight miles without issue, but now experienced dyspnea on exertion at three to four miles with new nocturnal palpitations. The patient had no personal or family history of structural heart disease and has not lived outside of the U.S. for any extended period with the exception of one four-month deployment to the Middle East. Medical history revealed only a history of pediatric recurrent Streptococcus pyogenes pharyngitis as a child though no previously valvulopathy, autoimmune conditions, or recent streptococcal infections. He denied tobacco use or any recreational drugs. His

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vital signs on presentation included a heart rate of 60 beats per minute, a blood pressure of 138/78mm Hg, a respiratory rate of 14 breaths per minute, with an oxygen saturation of 100% on room air, and an oral temperature of 97.9°F. His cardiovascular exam revealed a regular rate and rhythm with a grade 3 systolic ejection murmur at the left upper sternal border, which radiated to the axilla, though no lower extremity edema or rales was found on physical exam. Initial electrocardiogram (ECG) demonstrated sinus rhythm with benign early repolarization and intraventricular conduction delay (Figure 1). A transthoracic echocardiogram (TTE) revealed severe eccentric, posterior mitral valve regurgitation with appropriate left ventricular contractility, though there was noted left ventricular enlargement (Figure 2).

FIGURE 1 The patient's electrocardiogram (ECG) following a symptomatic period demonstrating sinus bradycardia at a rate of 44 beats per minute, normal PR and QT intervals, QRS interval of 113 milliseconds, moderate intraventricular conduction delay, and benign early repolarization.

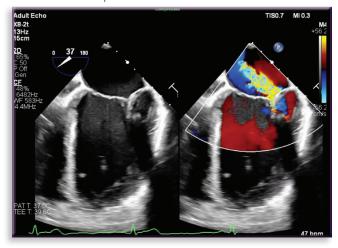


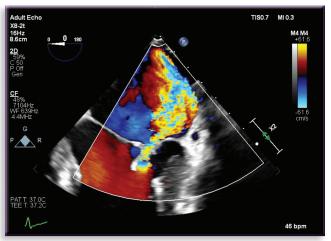
The patient was referred to cardiology, where a transesophageal echocardiogram (TEE) redemonstrated severe MR with a posterior regurgitant fraction of 27% and rheumatic appearing mitral valve leaflets. An ambulatory cardiac monitor was otherwise unremarkable without tachydysrhythmia. The patient was referred to cardiothoracic surgery, where a successful, mechanical mitral valve replacement was performed via minimally invasive right thoracotomy. He was successfully started on anticoagulation with a lifelong international normalized ratio (INR) goal of 2-3 and completed cardiac rehabilitation without issue. Repeat post-operative TTE demonstrated only trace prosthetic mitral valve regurgitation with normal left ventricular ejection fraction. The patient has returned to full cardiovascular exercise capacity and has been referred to the medical evaluation board.

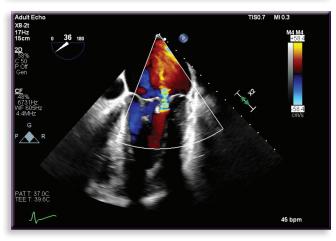
Discussion

RHD is a systemic, chronic post-infectious immune sequela of ARF. The initial insult most commonly occurs in childhood as beta-hemolytic group A streptococcal (GAS) pharyngitis. Primary screening is typically performed through use of the modified Centor score, rapid strep antigen testing, and/or throat culture. Suspected or confirmed infections are historically treated with a penicillin antibiotic as primary prevention for ARF.7 If patients fail to present for medical care or infections go untreated, the immune system generates cross-reactive antibodies to a streptococcal protein, causing it to attack its native endothelial cells, which results in widespread inflammatory damage.8,9 Consequently, ARF may occur weeks to months after the initial GAS pharyngitis infection. ARF can

FIGURE 2 The patient's transesophageal echocardiogram (TEE) showing severe posterior mitral valve regurgitation into the left atrium and severe left ventricular dilation.







be characterized by the Jones criteria, and diagnosis may be met by a combination of major and/or minor criteria, with the most common symptoms being fever and polyarthritis.⁵ Despite resolution of ARF, systemic immune response persists. While most endothelial cells possess regenerative capacity, those lining cardiac valves cannot be effectively repaired.^{5,9} This immune process remains progressive for several decades and leads to valvular fibrosis. The mitral valve is most significantly affected and may cause MS or MR resulting in mixed hemodynamic effects.¹⁰ Consequently, after decades of unchecked valvular dysfunction, chronic RHD will occur. The

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tricuspid or pulmonic valves are less frequently affected in RHD, and the aortic valve is only affected in <2% of cases.¹¹

Most patients who develop RHD will present with symptoms associated with mitral valve dysfunction, including shortness of breath, palpitations, fatigue, decreased exercise tolerance, and syncope or near-syncope.4 Worsening MS may cause increased left atrial pressures and lead to symptoms of heart failure such as orthopnea, paroxysmal nocturnal dyspnea, dysphagia, and dyspnea on exertion. 12 Severe valvular disease may also pose risk for exertional syncope and sudden cardiac death.¹² Physical exam may reveal murmurs, such as a mid-diastolic rumbling murmur heard in MS or a systolic murmur radiating to the axilla heard in MR.13 ECG changes are non-specific but may include p-wave changes, right-axis deviation, right ventricular hypertrophy, or arrhythmias such as atrial fibrillation in those with more severe mitral valve damage. 13 Auscultated murmurs should be investigated with TTE which may reveal valvular thickening, deformity, or decreased leaflet mobility. Referrals to cardiology and potentially cardiothoracic surgery should be placed.

Once congenital, acquired, or degenerative heart disease as an underlying cause are excluded by cardiology, a presumed diagnosis of RHD can be formally made using the 2012 World Health Federation criteria (Box 1).11 The criteria are based on age and evidence of echocardiographic findings. 10 For those under the age of 20 years, definite RHD criteria can be met by having both pathologic and morphologic RHD features within the mitral or aortic valves, or an MS gradient >4 mmHg.¹⁰ Borderline RHD criteria are met by having at least two morphologic features in the mitral valve with pathologic findings, or pathologic findings alone. For those over the age of 20 years, definite RHD is met in the same manner; however, there are no borderline criteria for this age group.

Once identified, initial management may begin with secondary antibiotic prophylaxis with a prolonged course of penicillin to eradicate the GAS infection, prevent disease progression, and decrease morbidity.14 Exercise restrictions and other lifestyle modifications may be implemented based on disease severity. Initial medical management for MS may be offered to control consequences of long-standing stenosis or regurgitation.15 Heart failure may be managed through goal-directed medical therapy with diuretics, angiotensin-converting enzyme inhibition (ACE-i) or angiotensin II receptor blocker (ARBs), beta-blockers, or mineralocorticoid receptor antagonists. 4,11,15 Atrial fibrillation can lead to thromboembolism, stroke, or worsening heart failure and should be managed with calcium channel blockers, beta-blockers, or digoxin with consideration for anti-coagulation. 4,15 Definitive management for severe disease may be achieved through percutaneous or surgical intervention via valvular repair or replacement. 4,15 Valvular repair can be performed via balloon mitral valvuloplasty or mitral commissurotomy and does not require definitive anticoagulation. 15 Valve replacements can be bioprosthetic or mechanical, though in contrast, mechanical valves require lifelong anticoagulation.15

RHD is a leading cause of preventable death and disability in children and young adults worldwide.1 Numerous recent global health initiations have increased primary screening and prevention, though RHD remains a significant burden across developing nations globally with an increased prevalence of BOX 1 2012 World Federation Criteria for Echocardiographic Diagnosis of RHD

For Age < 20 Years

Definite RHD (either A, B, C, or D):

- A) Pathological MR and at least 2 morphologic features of RHD of the MV
- B) MS gradient >4 mmHg
- C) Pathological AR and at least 2 morphologic features of RHD of the AV
- D) Borderline disease of both AV and MV

Borderline RHD (either A, B, or C):

- A) At least two morphological features of RHD of the MV without pathological MR or MS
- B) Pathological MR
- C) Pathological AR

Normal echocardiographic findings (either A, B, C, or D):

- A) Physiologic MR that does not meet all four Doppler echocardiographic criteria
- B) Physiologic AR that does not meet all four Doppler echocardiographic criteria
- C) Isolated morphological feature of RHD of the MV, without pathological MS or MR
- D) Morphological feature of RHD of the AV, without pathological AS or AR

For Age > 20 Years

Definite RHD (either A, B, C, or D):

- A) Pathological MR and at least two morphologic features of RHD of the MV
- B) MS gradient >4 mmHg
- C) Pathological AR and at least 2 morphologic features of RHD of
- D) Pathological AR and at least 2 morphologic features of RHD of

RHD = rheumatic heart disease; MR = mitral regurgitation; MV = mitral valve; MS = mitral stenosis; AV = aortic valve; AR = aortic regurgitation; AS = aortic stenosis.

RHD-related heart failure from 1990 to 2015.5 Once thought to be a declining disease process in the United States and only a result of foreign exposure, RHD has had an increase in age-adjusted mortality rates from 2017 to 2020 with projections for a surge in RHD-associated complications in the coming decade.3 Additionally, in a 10-year multicenter review by de Loizaga et al., 87% of diagnosed RHD cases had no travel to endemic regions, indicating a continued domestic burden of ARF and RHD.² Many patients and providers alike are unaware of the consequences of untreated strep throat infections, the overall disease burden, and increasing mortality rates. This case emphasizes the importance of primary screening and prevention in the U.S. for GAS infections, but also the need for increased echocardiographic evaluations and secondary prophylaxis when ARF is suspected in order to prevent progression to RHD.

The underlying physiologic effects and treatment of RHD present a risk of debilitation for military servicemembers, especially SOF servicemembers. While not specifically addressed in the Army Regulation for Standards of Medical Fitness, AR 40-501, consequences of RHD such as valvular dysfunction, valvular heart disease, atrial fibrillation, and long-term use of anticoagulants do not meet standards for military service. 16 Therefore, a Soldier with underlying valvular disease would be placed on a permanent profile and be referred to the Disability

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Evaluation System (DES) to determine if they are amenable for continued military service.¹⁶ Most will undergo a medical evaluation board process with subsequent medical retirement.

Conclusion

RHD is rare within the SOF population but may cause significant morbidity and potential mortality if unidentified and untreated. While recognition and treatment of streptococcal infection attempt to combat this disease process, autoimmune destruction can generate MS or MR. Patients present with chronic and progressive dyspnea, orthopnea, syncope or near-syncope, and exercise intolerance; in the SOF population, decreased exercise tolerance from their typically high baseline of physical fitness may be particularly noticeable. A thorough workup may demonstrate non-specific ECG changes and potential dysrhythmia, though diagnosis is confirmed with a TTE. Heart failure can be managed conservatively while the severity of mitral valvular disease will dictate if further intervention is indicated. SOF medics and providers should maintain a high level of clinical suspicion in those complaining of dyspnea, near-syncope or syncope, and decreased exercise tolerance to ensure timely diagnosis and treatment of this rare but potentially life-threatening condition.

Author Contributions

KO, DD, AC, and RB generated the manuscript. MM provided care for the patient and provided edits. All authors approved the final draft.

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