

VALVULAR HEART DISEASE

CASE REPORT: CLINICAL CASE

Lifelong Rheumatic Heart Disease Evolution and Need for Tailored Care



Sharona Hariri, MD, Kaylin T. Nguyen, MD, Joseph Hanna, MD, Jonathan Soverow, MD, Robin Wachsner, MD

ABSTRACT

Rheumatic heart disease leads to significant cardiovascular morbidity and mortality globally. This case presentation emphasizes the importance of considering patients' unique socioeconomic factors, such as crowded living conditions and multigenerational housing, to implement culturally sensitive approaches to prevention, diagnosis, secondary antibiotic prophylaxis, and management. (JACC Case Rep. 2024;29:102630) 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/>).

HISTORY OF PRESENTATION

This presentation follows the clinical course of a woman with rheumatic heart disease (RHD) over 27 years (Figure 1). The patient was 35 years old when a murmur was auscultated during pregnancy. A postpartum transthoracic echocardiogram (TTE) revealed rheumatic valvular disease involving the mitral and aortic valves, with severe mitral stenosis (MS), a mitral valve area (MVA) of 0.9 cm², moderate mitral regurgitation (MR), mild aortic stenosis, and mild aortic regurgitation.

Secondary antibiotic prophylaxis with intramuscular penicillin was initiated. One year later, she was

hospitalized with recurrent acute rheumatic fever (ARF), and the workup revealed elevated anti-streptolysin antibodies and a positive β -hemolytic streptococcus group A throat culture. She was noted to have variable compliance with oral antibiotic prophylaxis and her MS had progressed with an MVA of 0.77 cm². The Wilkins score was 5/16, and the patient underwent successful percutaneous mitral balloon valvuloplasty at age 37 years. Postprocedure, her MVA was 2.2 cm², and she remained in sinus rhythm for 3 years post-valvuloplasty, after which she had documented atrial fibrillation (AF), and warfarin was initiated. She subsequently had multiple additional admissions for acute decompensated heart failure and AF with rapid ventricular response, requiring multiple rate and rhythm control medications. She was continued on guideline-based antibiotic prophylaxis until she reached 40 years of age.

Seven years after valvuloplasty, at age 43 years, she presented with fever, chills, night sweats, and diffuse joint pain. She denied exposure to sick contacts or recent travel outside of the country. Vital signs were within normal limits. Physical

LEARNING OBJECTIVES

- To discuss the progressive course of rheumatic heart disease and significant cardiovascular morbidity.
- To identify social factors to consider in determining secondary antibiotic prophylaxis duration.

From the Division of Cardiology, University of California-Los Angeles, Olive View Medical Center, Sylmar, California, USA. The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the [Author Center](#).

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**ABBREVIATIONS
AND ACRONYMS**

- AF** = atrial fibrillation
ARF = acute rheumatic fever
MR = mitral regurgitation
MS = mitral stenosis
MVA = mitral valve area
PASP = pulmonary artery systolic pressure
RHD = rheumatic heart disease
TTE = transthoracic echocardiogram

examination was notable for prominent S₁, normal S₂, and grade II/IV diastolic and grade II/VI systolic murmur. She was admitted for parenteral antibiotics.

MEDICAL HISTORY

The patient's medical history was pertinent for hypertension and hypothyroidism. Social history was notable for emigrating from South America. She lived in a multigenerational home with her husband, children, and grandchildren. At the time of the initial MS diagnosis, her household included an adult child, 2 elementary school-aged children, and a newborn in addition to her and her husband. Within 1 year of her

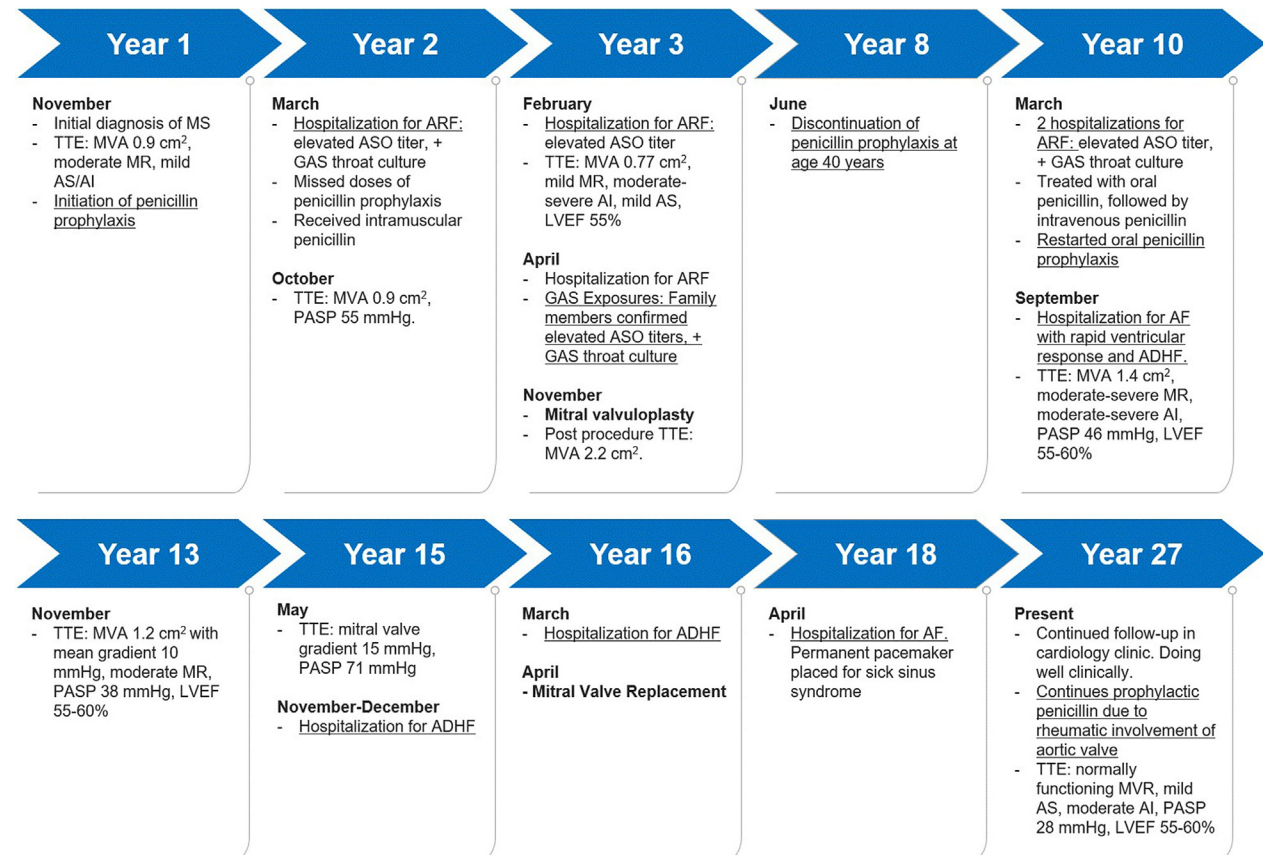
diagnosis, her child's partner and a grandchild were also living in the home. In total, there were up to 8 people, including 4 young children, living in a 1-bedroom apartment during this period.

DIFFERENTIAL DIAGNOSIS

Recurrent ARF, acute decompensated heart failure, acute renal failure, and infectious etiologies were all considered in the differential diagnosis.

INVESTIGATIONS

Laboratory evaluation revealed elevated anti-streptolysin antibodies of 1,408 IU/mL and positive β-hemolytic streptococcus group A throat culture. The patient's family members were noted to be positive

FIGURE 1 Timeline Displaying Clinical Progression of RHD as Related to Our Case Presentation

ADHF = acute decompensated heart failure; ARF = acute rheumatic fever; ASO = anti-streptolysin O; AF = atrial fibrillation; AI = aortic insufficiency; AS = aortic stenosis; LVEF = left ventricular ejection fraction; MR = mitral regurgitation; MS = mitral stenosis; MVA = mitral valve area; MVR = mitral valve replacement; PASP = pulmonary artery systolic pressure; GAS = group A streptococcus; TTE = transthoracic echocardiogram.

carriers for streptococcus infection on throat culture. TTE revealed a left ventricular ejection fraction of 60% to 65%, MVA of 2.2 cm², mild to moderate MR, moderate to severe aortic regurgitation, and pulmonary artery systolic pressure (PASP) of 42 mm Hg.

MANAGEMENT

Given recurrent presentations for ARF, high-transmission living environment, and variable compliance with oral therapy, the patient was transitioned to a monthly intramuscular penicillin injection. Five years later, a surveillance TTE revealed an elevated mitral valve gradient to 15 mm Hg and PASP of 71 mm Hg (Figures 2 to 4, Video 1).

She had multiple repeat hospitalizations for decompensated heart failure and ultimately underwent mitral valve replacement 13 years after her valvuloplasty at age 50 years. Two years later, she developed worsening paroxysmal AF and sick sinus syndrome, necessitating permanent pacemaker placement.

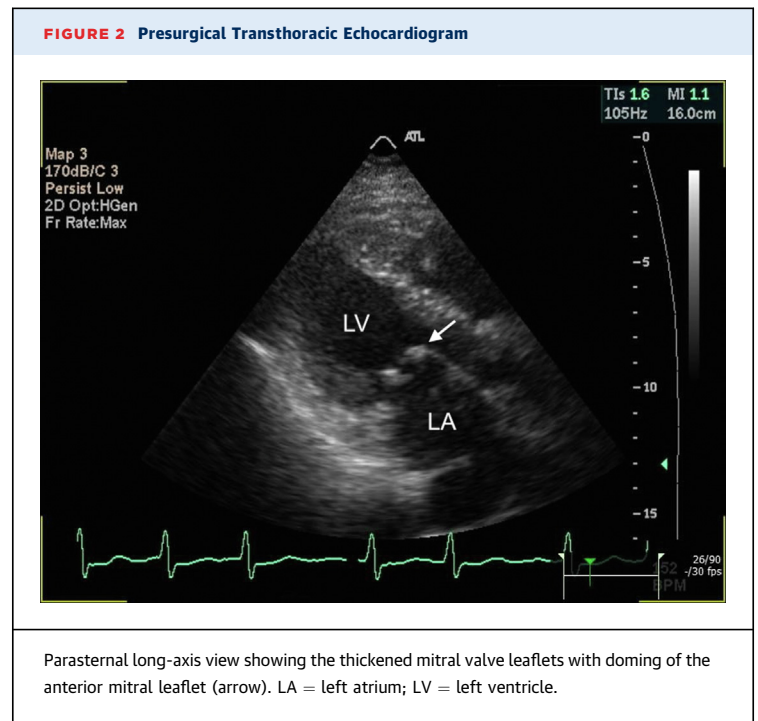
DISCUSSION

ARF and RHD affect an estimated 40 million people globally and are leading causes of cardiovascular death during the first 5 decades of life, prominent among marginalized populations.¹⁻³ In nonendemic regions such as the United States, mortality from RHD disproportionately affects populations of immigrants and those from low socioeconomic backgrounds.⁴

RHD more commonly affects the left-sided valves, particularly the mitral valve, causing acute valvulitis and valvular regurgitation. Isolated aortic disease can also occur but is rare.⁴ Chronic inflammation, progressive fibrosis, and recurrent bouts lead to commissural fusion and valvular stenosis. Patients with RHD can develop additional complications such as heart failure, endocarditis, arrhythmias, and cardiovascular death.⁴

Patients with a history of ARF are at very high risk of developing recurrences after subsequent group A streptococcal pharyngitis, which can lead to RHD or worsen existing RHD. Secondary prophylaxis with penicillin reduces streptococcal infections, recurrent attacks of ARF, and frequency and duration of hospitalization in patients with ARF or RHD.^{5,6} A recent randomized controlled trial of secondary antibiotic prophylaxis in Ugandan children and adolescents with latent RHD found that secondary antibiotic prophylaxis reduced the risk of disease progression at 2 years.⁷

Current guidelines are based primarily on expert opinion and lack a concise strategy for optimal



duration of antibiotic prophylaxis.² These recommendations focus on patient age, presence and severity of cardiac sequela, and time since the last episode of ARF. However, basing the duration of

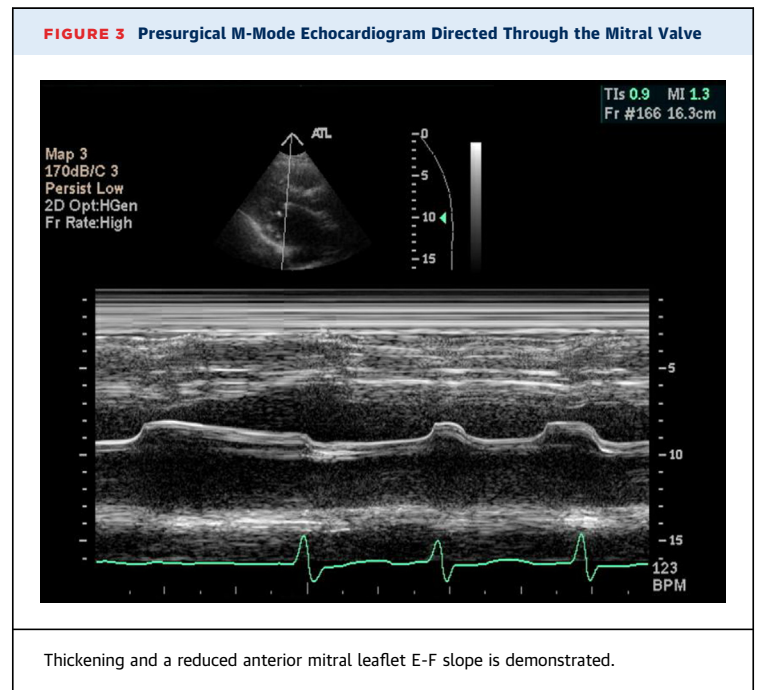
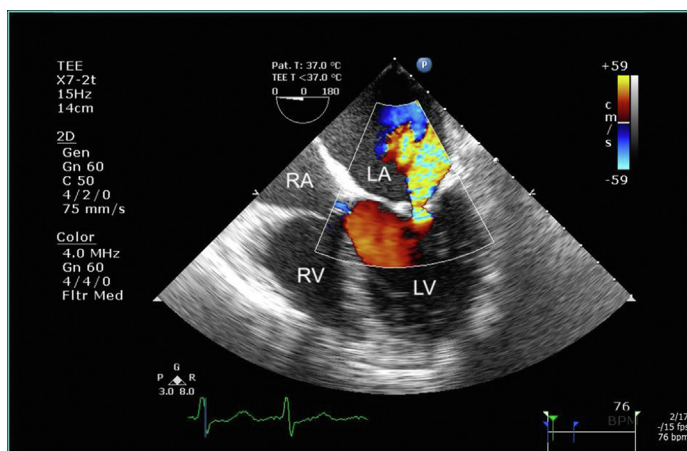
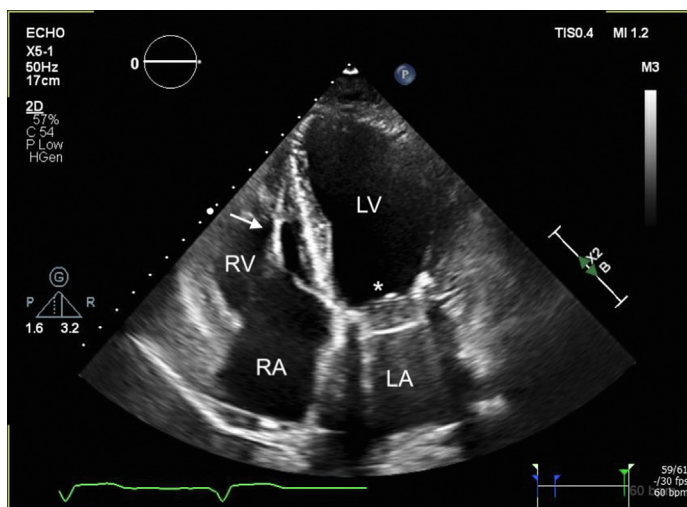


FIGURE 4 Presurgical Transesophageal Echocardiogram

Display of 4-chamber view with color Doppler demonstrating moderate mitral regurgitation. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

secondary antibiotic prophylaxis on the last episode of ARF can be problematic, as up to one-third of patients with ARF report no history of pharyngitis and most individuals with RHD have no known history of ARF.⁴ Current expert consensus recommends considering the risk of group A streptococcus

FIGURE 5 Transthoracic Echocardiogram

Display of 4-chamber view at 10 years after mitral valve replacement. Arrow demonstrates permanent pacemaker lead. Abbreviations as in Figure 4.

exposure, including high-risk occupations and living environments, before stopping prophylaxis. Moreover, in some individuals, lifetime prophylaxis may be necessary. Social risk factors such as residing within crowded areas, including multigenerational households, is common within some immigrant populations.² The number of multigenerational Latino households has been found to be about one-third or 31.7%.⁸ Additionally, high-risk status such as parents and grandparents with young children, healthcare workers, military recruits, and teachers should be considered.²

FOLLOW-UP

The patient has continued routine follow-up in the cardiology clinic and is overall doing well. Her most recent TTE was notable for normal left ventricle ejection fraction, normally functioning mechanical mitral valve, and a PASP of 28 mm Hg (Figure 5). She continues on metoprolol tartrate, amiodarone, digoxin, warfarin for AF, maintenance diuretic with furosemide, and prophylactic penicillin because of the rheumatic involvement of the aortic valve.

CONCLUSIONS

There is a strong need to highlight factors beyond patient age and time since the last ARF episode in considering the duration of secondary prophylaxis. Our case highlights the importance of consideration of socioeconomic, environmental, and cultural factors in the management of ARF and RHD within immigrant communities. Although the prevailing wisdom is that ARF is rare in patients over age 30 years, this does not appropriately weigh exposure through living and working conditions. By recognizing the unique circumstances of immigrant and underserved communities, physicians can implement culturally sensitive approaches to management, helping to mitigate the burden of RHD and improve health outcomes for vulnerable patient populations.

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ADDRESS FOR CORRESPONDENCE: Dr Kaylin T. Nguyen, UCLA-Olive View Medical Center, Department of Cardiology, Suite 2C-121, 14445 Olive View Drive, Sylmar, California 91342, USA. E-mail: knnguyen4@dhs.lacounty.gov.

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KEY WORDS rheumatic heart disease, secondary antibiotic prophylaxis

 **APPENDIX** For a supplemental video, please see the online version of this article.