

MULTIVALVULAR DISEASE

CLINICAL CASE

Predominant Rheumatic Tricuspid Stenosis



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ABSTRACT

BACKGROUND Diagnosis and management of multivalve disease could be challenging for clinicians. Rheumatic heart disease is a well-known etiology of multivalve disease.

CASE SUMMARY A 51-year-old male with a history of rheumatic heart disease was referred to rule out infectious endocarditis. Three-dimensional (3D) transesophageal echocardiography (TEE) showed significant stenosis of the tricuspid valve, moderate tricuspid regurgitation, rheumatic mitral valve disease, and a bicuspid aortic valve.

DISCUSSION Transthoracic echocardiography is the modality of choice for evaluation of cardiac valves and quantification of cardiac chambers. However, due to the complex nature of multivalve disease, using a complementary imaging technique such as 3D TEE becomes crucial in many patients. Tricuspid stenosis as the dominant valvular lesion in rheumatic heart disease is rare and almost always occurs in the presence of mitral valve disease. We present a unique case of rheumatic heart disease with the involvement of mitral and tricuspid valves in the presence of a bicuspid aortic valve, in which tricuspid stenosis is the predominant lesion. We also discuss the important role of cardiac imaging and 3D TEE in patient decision making.

TAKE-HOME MESSAGES Tricuspid stenosis as the dominant valvular lesion in rheumatic heart disease is rare and almost always occurs in the presence of mitral valve disease. Transthoracic echocardiography remains the modality of choice for evaluation of the tricuspid valve. 3D TEE can play an important role in reaching a final management decision. (JACC Case Rep. 2025;30:104808) 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/>).

BACKGROUND

Multivalve disease presents a diagnostic challenge. In many patients, there is a predominant valve lesion, and symptoms and pathophysiology resemble those of a pure dominant lesion.¹ Rheumatic heart disease is a well-known etiology of multivalve disease, in which mitral valve involvement is almost

always present and predominant. Although tricuspid valve involvement is not uncommon, tricuspid stenosis (TS) as the predominant pathology is rare in rheumatic heart disease.^{2,3} We present a case of multivalve disease in which the predominant lesion is TS, with emphasis on the role of a multiparameter approach by echocardiography in the diagnosis and management of the patient.

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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

MR = mitral regurgitation
RA = right atrium/atrial
TEE = transesophageal echocardiography
TR = tricuspid regurgitation
TS = tricuspid stenosis
TTE = transthoracic echocardiography
3D = 3-dimensional

CASE SUMMARY

A 51-year-old man with a history of rheumatic heart disease presented with the complaint of linear discoloration on his fingernails consistent with splinter hemorrhage on physical exam. The patient had a history of Addison's disease and primary biliary cholangitis. Due to the history of rheumatic heart disease and the presence of splinter hemorrhages, infective endocarditis was considered the first diagnosis. Two sets of blood cultures were negative for bacteremia. Electrocardiography showed a sinus rhythm with evidence of biatrial enlargement. Transthoracic echocardiography (TTE) revealed a significant increase in the diastolic gradient at the tricuspid valve level: a mean gradient reported 9 mm Hg at a heart rate of 68 beats/min. No prior Echo study was available to compare. TTE was not conclusive in excluding infective endocarditis. The patient was referred for a transesophageal echocardiography (TEE).

A detailed morphologic study was performed using 2-dimensional and 3-dimensional (3D) TEE. The 3 tricuspid leaflets were thickened with no significant calcification. The leaflet's motion was severely restricted, demonstrating the typical doming often seen in rheumatic involvement. The commissure was thickened and fused, leading to a severely restricted opening. The subvalvular structures were shortened and retracted. The tricuspid valve orifice area was measured using a multiplanar 3D dataset for optimal alignment and revealed a tricuspid valve area $<3 \text{ cm}^2$ (Figure 1, Video 1). No vegetation or any other echocardiographic sign in favor of infective endocarditis was noted. The hemodynamic assessment of the

TAKE-HOME MESSAGES

- The complex nature of multivalvular disease can be a challenge for clinicians and requires comprehensive multiapproach imaging that involves anatomical and hemodynamic assessment of each valve and careful evaluation of cardiac chamber size and function.
- Mixed rheumatic heart disease is a common cause of multivalvular disease. Rheumatic TS occurs in concert with TR, rheumatic mitral valve disease, and/or aortic valve disease. It is uncommon for TS to be the predominant lesion in rheumatic heart disease.
- TEE with 3D application should be considered for detailed evaluation of the valve anatomy, precise hemodynamic evaluation, and decision-making process.

tricuspid valve was performed by spectral and color Doppler study. There was a significantly increased diastolic gradient at the tricuspid level (tricuspid valve velocity time integral: 70 cm; tricuspid valve peak/mean gradient: 15/5 mm Hg; tricuspid valve pressure half-time: 210 ms). There was moderate tricuspid regurgitation (TR). The TR peak gradient was measured as 17 mm Hg (Figure 2, Video 2).

The mitral valve exhibited leaflet thickening more dominant at the tip of the leaflets, leading to a doming and classic hockey stick appearance of the anterior leaflet, pathognomonic for rheumatic valve disease. The posterior leaflet and subvalvular structures were thickened and retracted. The medial and lateral commissures were mildly fused, causing the fish mouth opening of the mitral valve despite no significant increase in diastolic gradient (the mitral

VISUAL SUMMARY Multivalvular Heart Disease With Predominant Rheumatic Stenosis

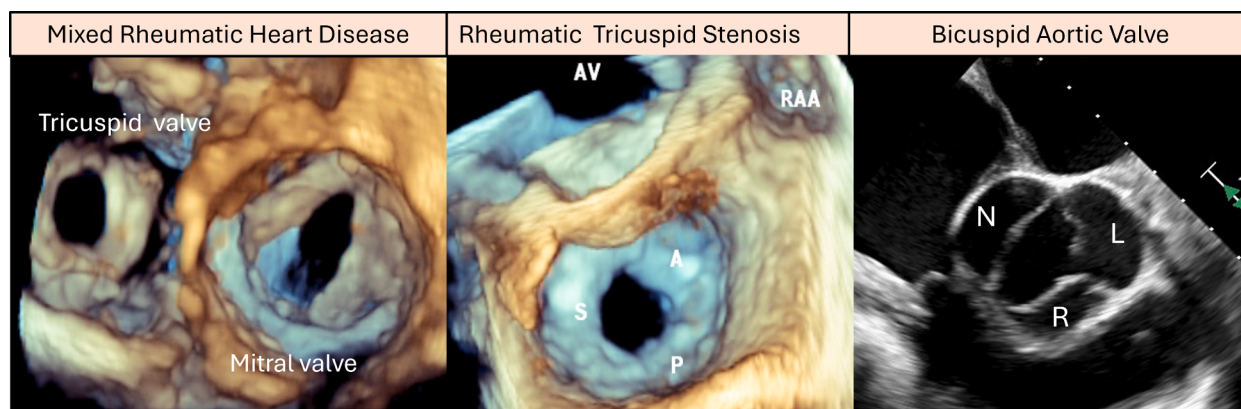
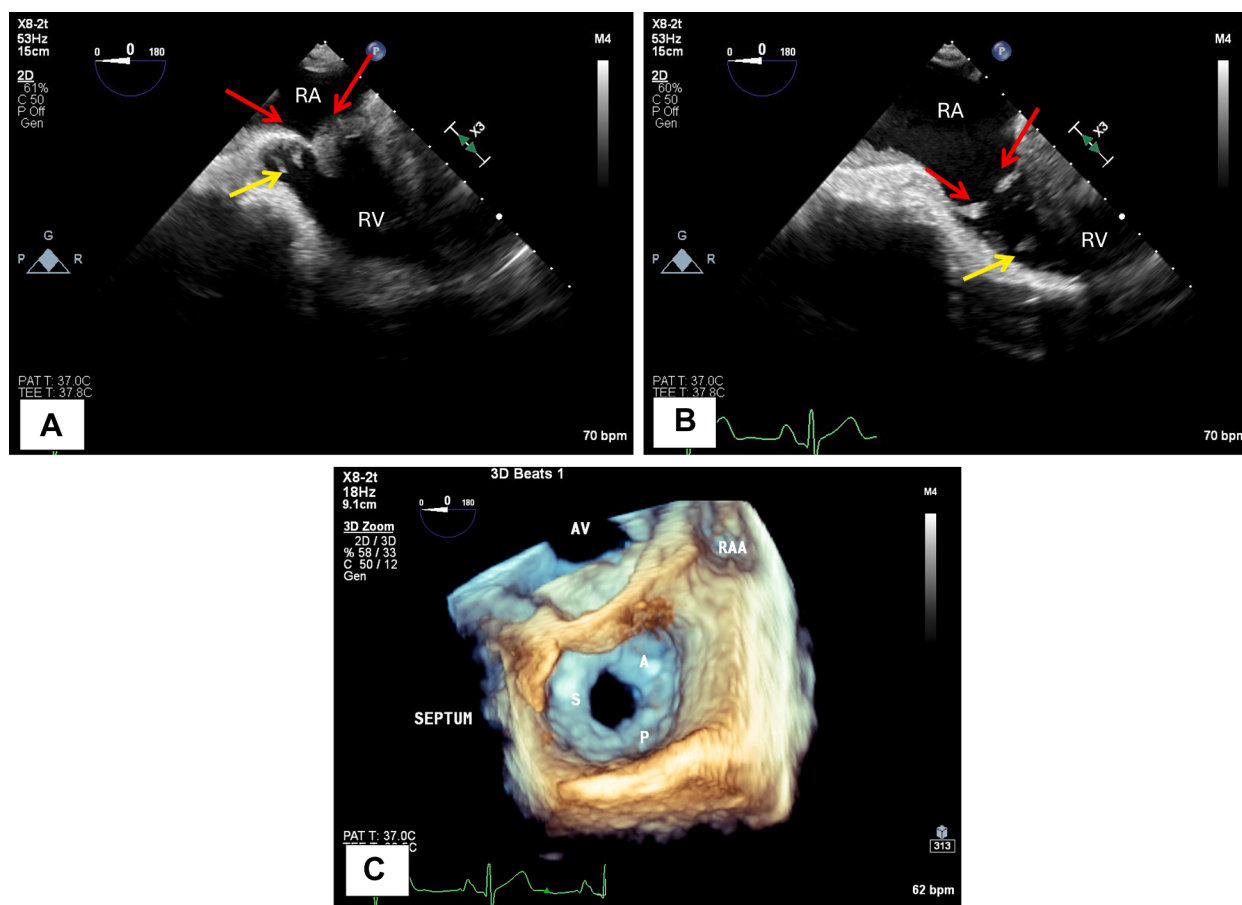


FIGURE 1 Morphologic Changes in Rheumatic Tricuspid Stenosis



(A, B) Two-dimensional and (C) 3-dimensional midesophageal transesophageal echocardiography views of the tricuspid valve illustrate morphologic findings of severe rheumatic stenosis. There are 3 leaflets: anterior (A), posterior (P), and septal (S), with thickening, fusion, and doming appearance (red arrows). Note the retraction and shortening of subvalvular structures pointed with yellow arrows. The commissural fusion results in a restricted fish mouth opening in diastole, illustrated in C and [Video 1](#). AV = aortic valve; RA = right atrium; RAA = right atrial appendage; RV = right ventricle.

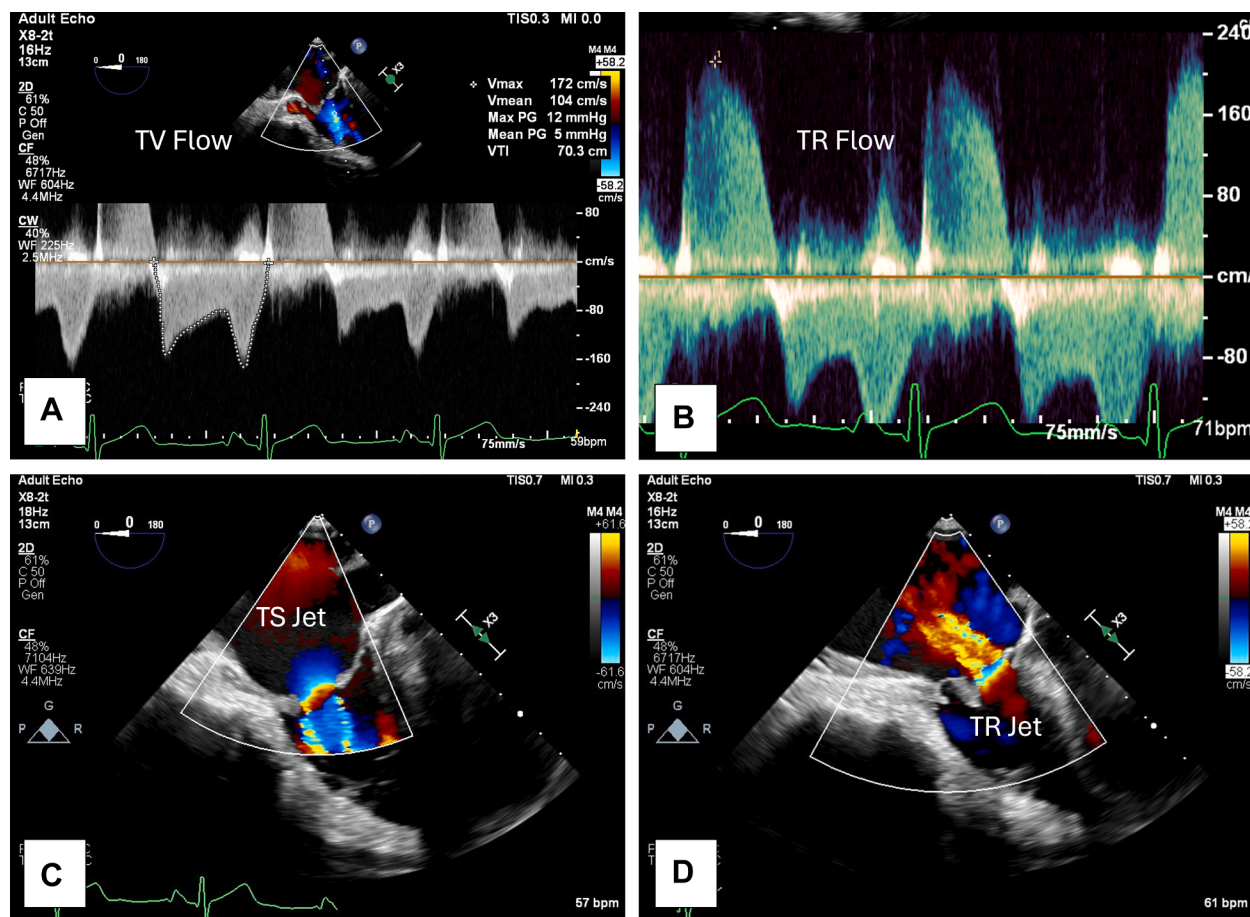
valve velocity time integral was 36 cm, and the mitral valve mean and peak gradient were measured as 3/7 mm Hg). There was moderate mitral regurgitation (MR) by Doppler study ([Figure 3](#), [Video 3](#)). The aortic valve was anatomically trileaflet, with a raphe between the right and left coronary cusps, leading to the functionally bicuspid aortic valve. The aortic valve did not exhibit the typical findings of rheumatic heart disease, such as thickening of the tip of the leaflets, commissural fusion, and triangular opening.⁴ There was mild aortic regurgitation and no aortic stenosis. The pulmonic valve was normal in morphology and function.

The left and right ventricles were normal in size and function. The size of the left atrium was at the upper limit of normal. The right atrium (RA) was

severely enlarged (RA area: 28 cm²; RA volume: 98 mL; body surface area: 1.8 m²). The right-sided heart chamber quantification and TR severity evaluation were done based on the reference limits recommended by the American Society of Echocardiography and the American College of Cardiology's newest guidelines.⁵ The interatrial septum appeared aneurysmal, with the septum bulging toward the left atrium ([Figure 4](#), [Video 4](#)). Contrast injection revealed no patent foramen ovale.

The patient was diagnosed with mixed rheumatic valvular disease of severe TS, moderate TR, and moderate MR with concomitant bicuspid aortic valve and mild aortic regurgitation. TS was deemed the predominant lesion, and the patient was approached accordingly. The valvular severity was categorized as

FIGURE 2 Hemodynamic Assessment of TV



(A, B) Spectral Doppler tracings and (C, D) color Doppler by transesophageal echocardiography were obtained from a modified 4-chamber midesophageal view. (A) Tricuspid stenosis; (B) tricuspid regurgitation. (C, D) Color Doppler recordings that demonstrate the accelerated and regurgitant jet through the tricuspid valve (TV) during diastole and systole, respectively. Tricuspid stenosis and tricuspid regurgitation (TR) are commonly seen concomitantly in rheumatic tricuspid involvement, as shown in our patient (corresponds to Video 2).

stage C1 based on the 2020 American College of Cardiology/American Heart Association guidelines for the management of patients with valvular heart disease.⁶ The patient elected for a medical follow-up and was advised to return for an echocardiography study in 6 months. No secondary prevention for rheumatic fever was suggested because the patient has received antibiotic prophylaxis for more than 10 years and has passed the 40-year-old cutoff.¹

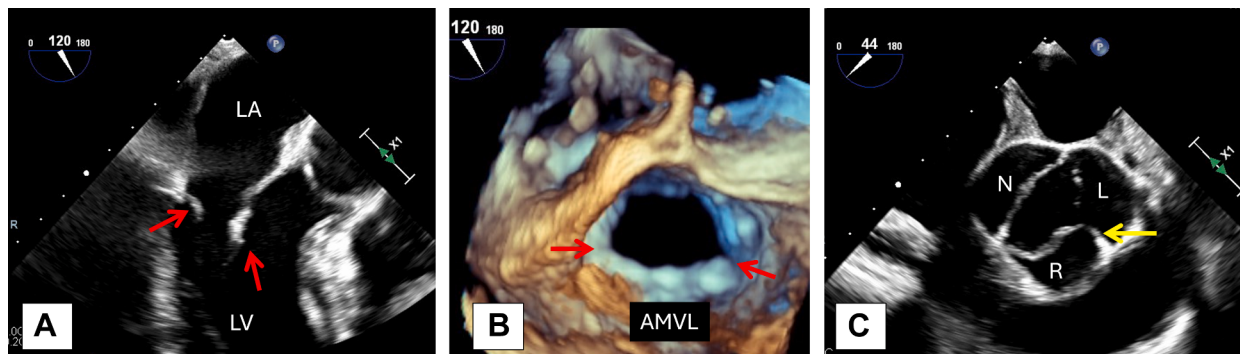
DISCUSSION

Rheumatic heart disease is one of the main etiologies of multivalve disease. Rheumatic involvement of right-sided valves is less common than their left-

sided counterparts and receives less attention. Rheumatic TS is a rare presentation of rheumatic heart disease and usually occurs in concert with TR, rheumatic mitral valve disease, and/or aortic valve disease. TS as the dominant manifestation of rheumatic heart disease, such as in our patient, is unusual. This case highlights the echocardiographic findings of dominant TS, rather than TR, mitral stenosis, or mitral regurgitation, which are more commonly encountered in rheumatic heart disease.

Echocardiography is the diagnostic tool of choice in the comprehensive assessment of the right heart.⁵ Whenever TTE is not conclusive, TEE with the application of 3D should be performed. The main goal of the echocardiography study is the evaluation of

FIGURE 3 Morphologic Changes of Mitral and Aortic Valves



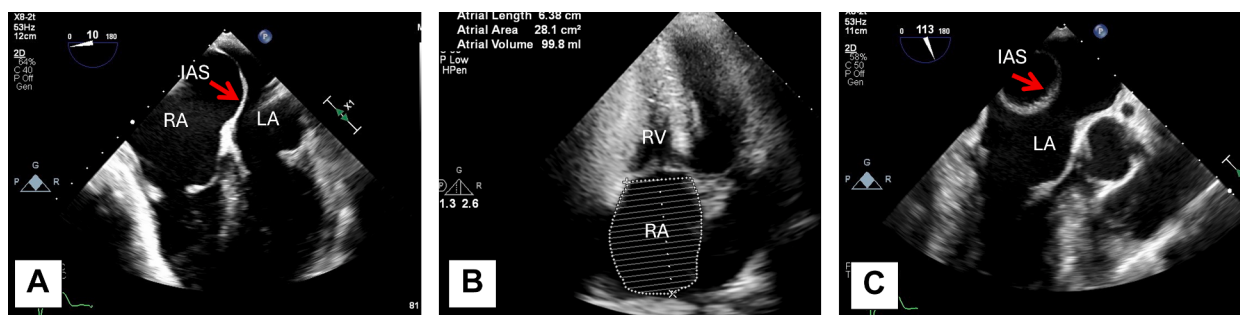
(A) Two-dimensional transesophageal echocardiography (TEE) midesophageal long-axis view in diastole demonstrates mitral leaflet thickening dominant at the tips leading to a hockey stick or bent-knee appearance (pointed with arrows). (B) Three-dimensional TEE zoomed-in image of the mitral valve; arrows point to commissural fusions, which are the hallmark of rheumatic mitral disease (corresponds to [Video 3](#)). (C) Two-dimensional TEE midesophageal short-axis view of the aortic valve demonstrates 3 anatomic leaflets with a raphe between the left and right cusps (pointed by yellow arrow). There is no leaflet thickening, scar tissue, or retraction in favor of rheumatic involvement. AMVL = anterior mitral valve leaflet; L = left coronary cusp; LA = left atrium; LV = left ventricle; N = noncoronary cusp; R = right coronary cusp.

the tricuspid valve anatomy, the severity of stenosis and regurgitation, and its hemodynamic effects on the other cardiac valves, heart chambers, and pulmonary artery pressure. The use of 3D TEE allows for “en face” imaging of the tricuspid valve and accurate measurement of annulus size and concomitant regurgitation.⁷

The morphological characteristics of rheumatic TS are very similar to those found in rheumatic mitral stenosis. However, significant calcification, commonly noted in mitral stenosis, is not seen in TS

to the same extent.⁷ Detailed anatomical evaluation should be performed with special attention to the leaflet morphology, mobility, and diastolic doming. The degree of leaflet thickening, calcification, and retraction should be examined, as well as commissural fusion, and shortening of the subvalvular apparatus. Our case did not have significant calcification of tricuspid leaflets, but the 3 leaflets demonstrated thickening and restricted motion, and the commissures were fused. 3D echocardiography allows visualization of the stenotic valve area from

FIGURE 4 RA Changes in Rheumatic Tricuspid Stenosis



(A, C) Two-dimensional transesophageal echocardiography midesophageal 4-chamber and modified long-axis views in diastole demonstrate bowing of the interatrial septum (IAS) (red arrow) toward the left atrium (LA) in favor of elevated right atrial (RA) pressure. Because the IAS is aneurysmal, the bowed part of the septum could mimic a mass in the atrium. (B) Two-dimensional transthoracic echocardiography modified apical 4-chamber view demonstrates severe RA enlargement due to chronically elevated RA pressure (see also [Video 4](#)). RV = right ventricle.

both atrial and ventricular perspectives and is particularly useful for deriving tricuspid valve area by multiplanar images of 3D datasets.³

Tricuspid valve involvement in rheumatic heart disease is commonly a combination of regurgitation and stenosis. In rare cases in which TS is the predominant lesion, progressive reduction in the tricuspid valve area results in increased RA pressure, increased diastolic pressure gradient between RA and right ventricle, and decreased right ventricular forward stroke volume. TS severity should be evaluated by spectral and color Doppler studies. TR severity should be evaluated comprehensively with qualitative and quantitative measurements.³ The presence of severe TR is reported as an independent predictor of heart failure and other major adverse cardiac events in multiple studies.²

The aneurysmal motion of the interatrial septum, bulging toward the left, represents increased RA pressure in our patient; however, no shunt was detected with contrast injection. The aneurysmal interatrial septum could be mistaken for a mass or clot in the atrium. Other echocardiographic findings of elevated RA pressure, which support the severity of TS, include hepatic vein congestion, plethoric inferior vena cava, and increased RA size; however, the diagnosis of primary biliary cholangitis in our patient reduced the specificity of these findings.

TS is almost always accompanied by some degree of rheumatic mitral and/or aortic valve disorder.³ Isolated rheumatic involvement of the tricuspid valve is rare. In the absence of rheumatic mitral valve disease, one should consider other etiologies of TS, such as tumors, masses, or calcifications.^{8,9} In our patient, the rheumatic etiology of TS was bolstered by typical findings of rheumatic disease in the mitral valve, including leaflet thickening, commissural fusion, and chordal shortening, which led to MR.

Although aortic regurgitation in the presence of rheumatic MR and TS implies rheumatic etiology as the first differential diagnosis, the echocardiography study of our patient did not reveal any of the findings typically seen in rheumatic heart disease. The aortic

valve possessed 3 thin leaflets with no scar, doming, or retraction. The commissure between the right and left cusps was fused, causing a functional bileaflet valve with eccentric opening.

Classification of valve disease severity is based on symptoms, valve anatomy, and the effect of the valve lesion on ventricular and vascular function. In multivalve disease, the difficulty lies in attributing symptoms to the valvular disease at hand, in our case, TS. It may be that neither of the lesions by themselves reaches stage C, yet in combination they may be severe enough to necessitate intervention, or different lesions can balance each other; in our case, this resulted in a more balanced picture and delay seeking therapy by the patient.⁶

CONCLUSIONS

The complex nature of multivalve disease can be a challenge and requires comprehensive multi-approach imaging that involves anatomical and hemodynamic assessment of each valve and careful evaluation of cardiac chamber size and function.

TTE is the primary modality of choice for anatomical and functional evaluation of cardiac valves in multivalve disease. 3D TEE plays an important role in reaching a final management decision. Limited recommendations are available for the management of mixed valvular disease with dominant TS (Video 5). Decisions are made based on clinical, electrocardiography, echocardiography, supplemental paraclinical findings, and patients' preferences.

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Dr Saric has served as a speaker for Abbott Laboratories, Boston Scientific, Medtronic, and Philips and on the advisory board of Siemens. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS bicuspid aortic valve, mitral valve, rheumatic heart disease, tricuspid valve

 **APPENDIX** For supplemental videos, please see the online version of this paper.