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VALVULAR HEART DISEASE

CASE REPORT: CLINICAL CASE

# Rheumatic Pulmonary Stenosis in an Elderly Man

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## ABSTRACT

An elderly man presented with right ventricular outflow obstruction due to severe pulmonary stenosis. Rheumatic changes were noted in mitral, aortic, and pulmonary valves. However, pulmonary valve was the predominantly affected valve. This is the first case report of rheumatic heart disease presenting predominantly with severe pulmonary stenosis in the elderly. (JACC Case Rep. 2024;29:102626) © 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/).

## HISTORY OF PRESENTATION

A 66-year-old man, from Morocco, came to Saudi Arabia as a visitor to perform a pilgrimage (Hajj). He presented to our institution with fatigue and dyspnea of NYHA functional class III. He denied orthopnea, paroxysmal nocturnal dyspnea, chest pain, palpitation, syncope, or presyncope. He reported frequent febrile illnesses during childhood but no history of diagnosed rheumatic fever. On examination, he was afebrile with a blood pressure of 161/72 mm Hg, pulse of 74 beats/min and regular, respiratory rate of 18 breaths/min, and oxygen saturation of 98% on room air. Auscultation revealed a loud S2 (P2), with a 3/6 ejection systolic murmur over the pulmonary area. Jugular venous pressure was 4 cm above the sternal angle with +1 bilateral lower limb, pitting edema. There was no organomegaly or signs of hepatic congestion.

### LEARNING OBJECTIVES

- To make a differential diagnosis of pulmonary stenosis presenting in the elderly.
- To be familiar with echocardiographic finding of rheumatic valvular disease including the rare involvement of right-sided valve including the PV.

## MEDICAL HISTORY

His symptoms dated back to 1 year before presentation, but he did not seek any medical attention in his country. He denied any chronic illness.

## DIFFERENTIAL DIAGNOSIS

Differential diagnoses included pulmonary stenosis, pulmonary embolism, and pulmonary hypertension.

## INVESTIGATIONS

Complete blood count, kidney function, cardiac markers, glycosylated hemoglobin, lipid profile, and liver enzymes were unremarkable. A 12-lead electrocardiogram showed sinus rhythm with right-axis deviation and inverted T-wave in the inferior leads (Figure 1). Transthoracic echocardiogram showed prominent thickening of the pulmonary valve (PV)

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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**

**PV** = pulmonary valve

**RV** = right ventricle

with commissural fusion and systolic doming (suggestive of rheumatic etiology). It was associated with very severe valvular pulmonary stenosis, peak velocity of 5.8 m/s, peak pressure gradient of 134 mm Hg (**Figures 2A and 2B**, Video 1), and mild pulmonary regurgitation (**Figure 3**). There was right ventricular hypertrophy and preserved right ventricle (RV) size and systolic function. The mitral valve leaflets and the chordae tendineae were thick, causing restricted movement of the anterior leaflet (hockey stick appearance) but minimal stenosis (calculated mitral valve area of 3.1 $cm^2$, pressure half-time of 117 cm/s, mean pressure gradient of 1 mm Hg) and mild mitral regurgitation (**Figures 4A and 4B**, Video 2). The aortic valve is thickened without stenosis, but mild aortic regurgitation (**Figures 5A to 5C**, Video 3). The left ventricle was normal in size and systolic function. The right atrium was dilated. Cardiac computed tomography confirmed the rheumatic changes of the mitral valve (restricted movement of the anterior leaflet due to thickening and the leaflet and subvalvular apparatus with commissural fusion) and aortic valve (thickening at the edge of the leaflets) (**Figure 6**, Videos 4 and 5). The PV was thickened with commissural fusion (**Figure 7A and 7B**, Videos 6 and 7). Right ventricular hypertrophy was noted (**Figure 8**). There was no subvalvular, supra-valvular obstruction, or other associated congenital anomalies. There was no obstructive coronary artery disease. Calcium score was 12.

## MANAGEMENT

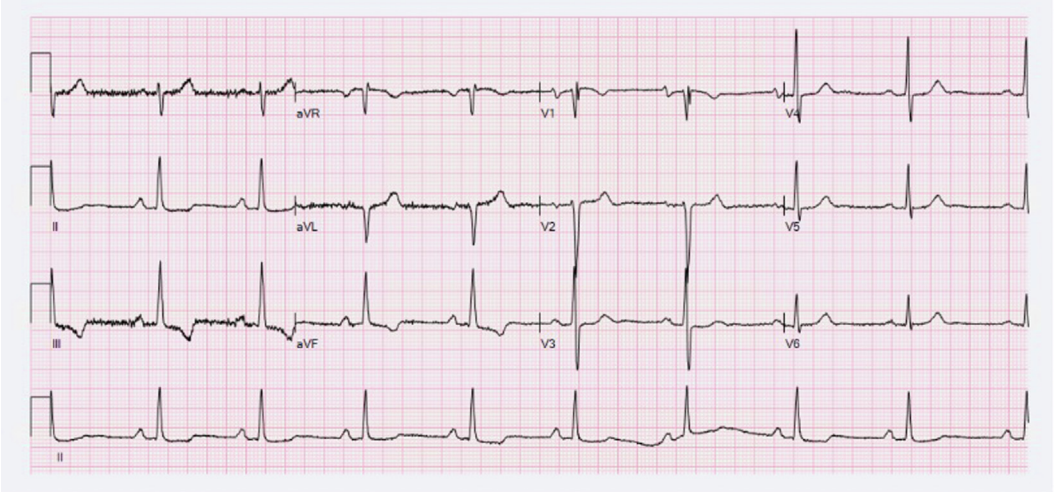
Symptomatic management with intravenous furosemide was started. We discussed the case in the structural heart team meeting. Given the absence of valvular dysplasia, supra-valvular and subvalvular obstruction, other valvular major dysfunction, and coronary artery disease, it was decided to perform pulmonary balloon valvuloplasty with a plan of sequential balloon inflation in the future to avoid RV suicide and acute pulmonary edema. It was performed using an 18-mm Z-Med balloon based on the annulus measurement of 18 mm on computed tomography.

Invasive hemodynamics post procedure, demonstrated a decrease in the peak transpulmonary gradient from 85 to 52 mm Hg, and right ventricular systolic pressure from 107 to 76 mm Hg.

## DISCUSSION

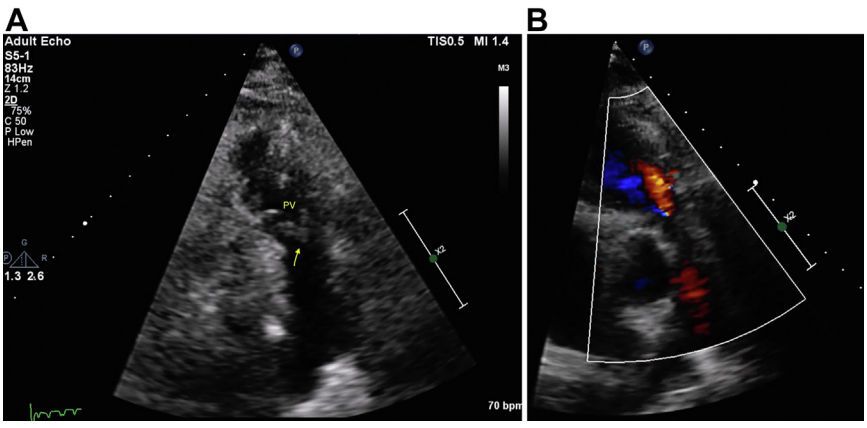
Pulmonary stenosis is the most common cause of the right ventricular outflow obstruction. It is mostly congenital and often associated with congenital

**FIGURE 1** 12-Lead Electrocardiogram




12-lead electrocardiogram shows right-axis deviation, inverted T-wave in the inferior leads.

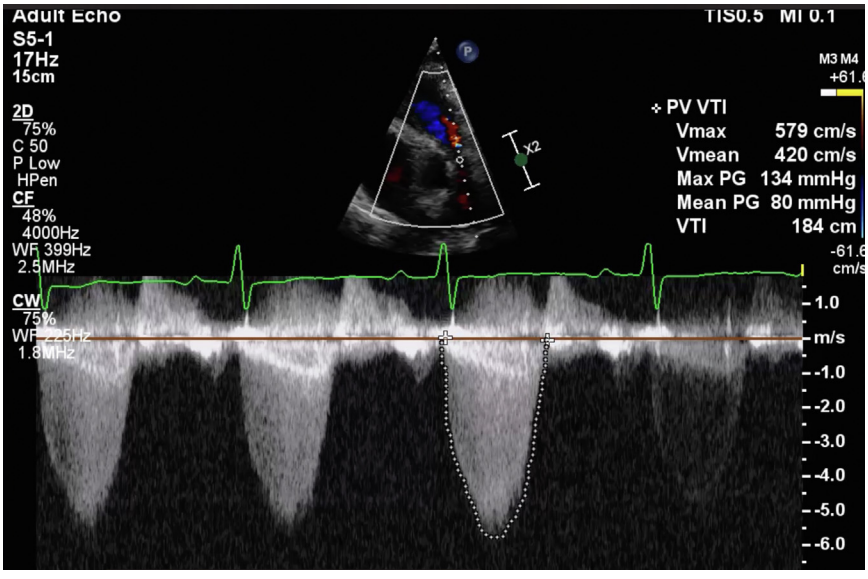
**FIGURE 2** **Parasternal Short-Axis View of the Pulmonary Valve**




Short-axis view showing (A) the commissural fusion of PV with systolic doming. (B) Color imaging showing mild pulmonary regurgitation. PV = pulmonary valve.

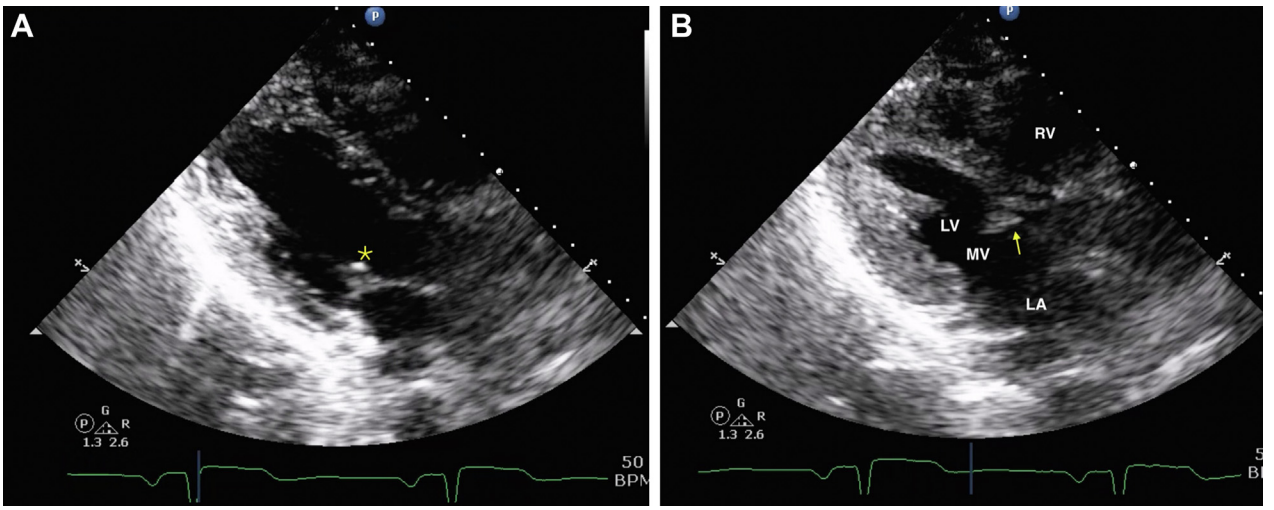
syndromes and other congenital heart disease, but can occur in isolation in 8% to 10%. It can be acquired from rheumatic heart disease, carcinoid, trauma, or after infective endocarditis.[1] Isolated rheumatic pulmonary valvular stenosis has been reported rarely in children from developing countries.[2] It usually presents in adulthood but is rarely diagnosed first in the elderly. We report a case of severe pulmonary stenosis presents in an elderly patient, posing a diagnostic challenge regarding the etiology. Buendía et al[3]

**FIGURE 3** **Continuous-Wave Doppler Imaging of the Pulmonary Valve**



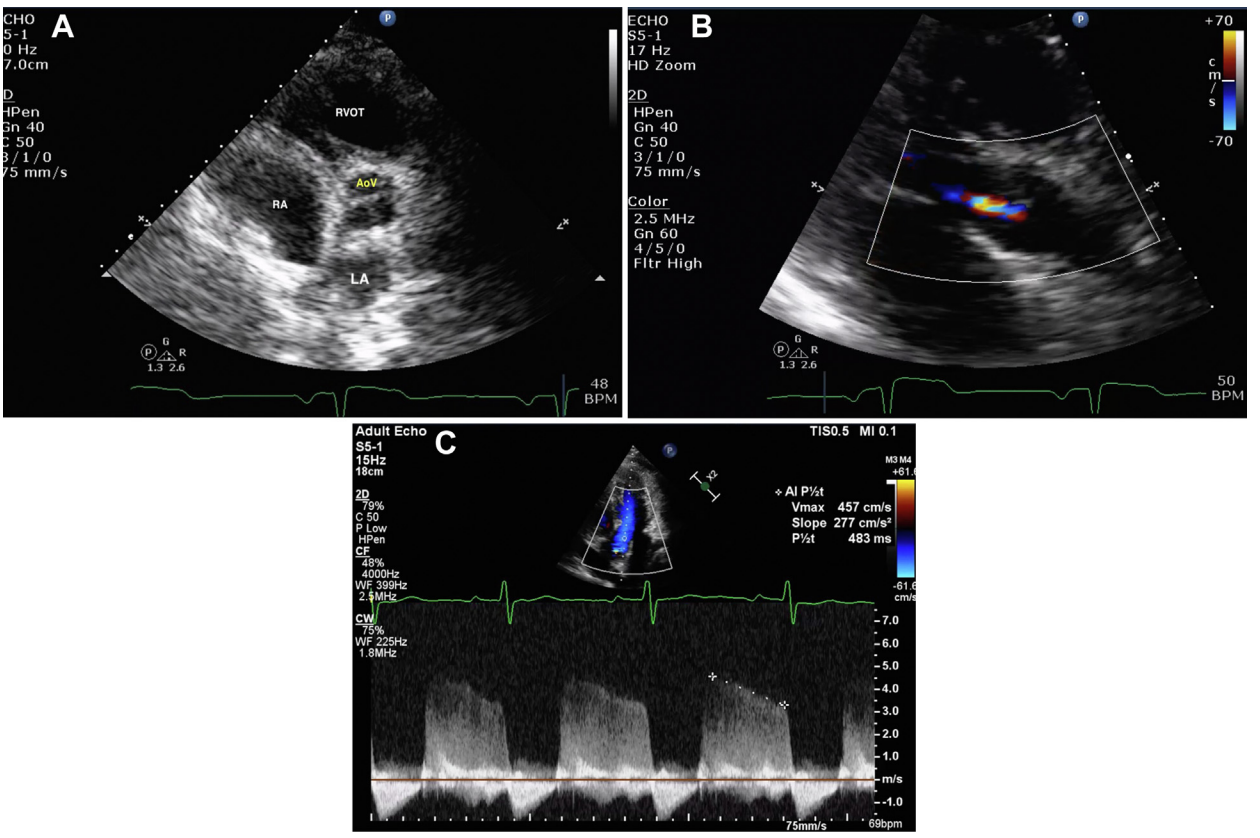

Continuous-wave Doppler imaging showing the velocities of pulmonary stenosis and regurgitation.

**FIGURE 4 Parasternal Long-Axis View of the Mitral Valve**



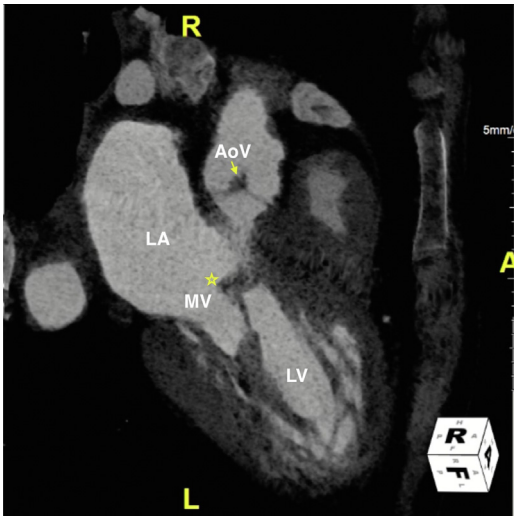

Parasternal long-axis view showing thickened leaflets and chordae tendineae with calcification of the chordae (*) in systole (A). Restricted anterior leaflet motion causing hockey stick appearance (arrow) (B). LA = left atrium; LV = left ventricle; MV = mitral valve; RV = right ventricle.

**FIGURE 5 Echocardiographic Assessment of the Aortic Valve**




(A) Parasternal short-axis view showing mild thickening of the cusps. (B) Color Doppler showing mild aortic regurgitation. (C) Continuous wave showing the velocities of aortic regurgitation with no significant stenosis. AoV = aortic valve; RA = right atrium; RVOT = right ventricular outflow tract; other abbreviations as in Figure 4.

**FIGURE 6** Cardiac Computed Tomography




Computed tomography, long-axis view showing thickened anterior leaflets and chordae tendineae with restricted movement of the anterior leaflet (hockey stick appearance, *), with thickening of the aortic valve at the margin (arrow), suggestive of rheumatic changes of both valves. Abbreviations as in Figures 4 and 5.

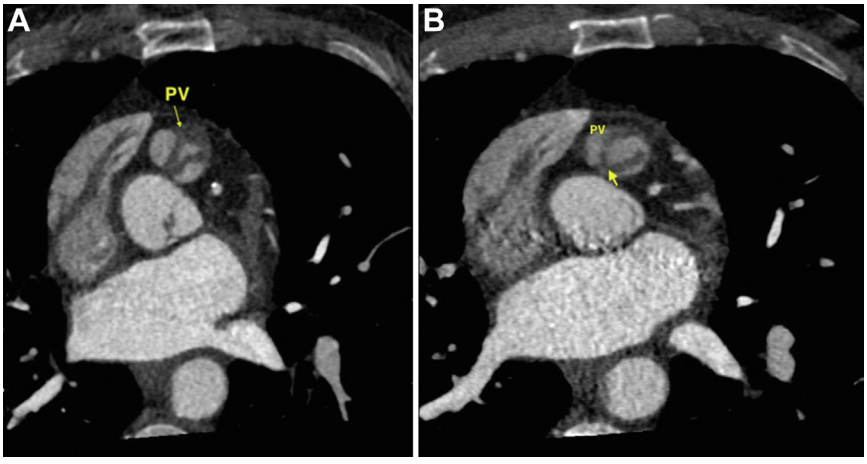
reviewed 100 postmortems of patients with rheumatic involvement of the PV. Isolated PV involvement was found in 0% of cases, but was reported in significantly greater incidence of rheumatic pulmonary disease in patients with coexisting congenital lesions of the valve. In our case, coming from a developing country and the typical rheumatic changes seen in the pulmonary, mitral, and aortic valves make the rheumatic etiology most favorable. Predominance of the pulmonary involvement can be caused by coexisting congenital lesion of the valve, but this cannot be confirmed without surgical exploration of the valve.

Rheumatic pulmonary stenosis is rare, and a scoring system for the suitability of balloon valvuloplasty is not established in the literature. Guidelines for the management of pulmonary stenosis were followed, which are based on congenital pulmonary stenosis, recommending percutaneous balloon valvuloplasty in the absence of other surgical indications as the first choice in management.[4,5]

## FOLLOW-UP

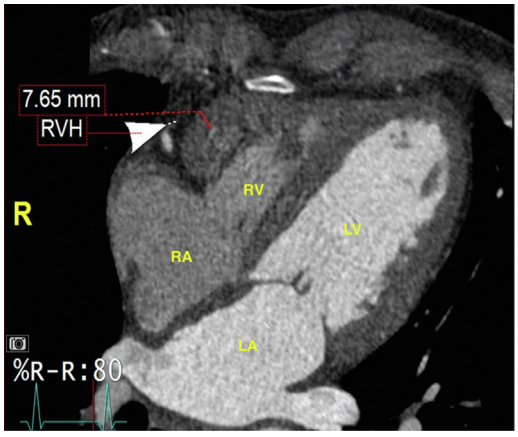
The patient tolerated the procedure well. Follow-up echo, done the next day, confirmed the reduction of trans-PV gradient from 100 mm Hg to 41 mm Hg with mild regurgitation noted (**Figure 9**, Video 8). He was discharged to complete his pilgrimage and he returned back to his home country. He was advised to follow up with a cardiologist for clinical assessment, repeat electrocardiogram, and echo within 6-8 weeks, and then yearly for the first year.

**FIGURE 7** Cardiac Computed Tomography




Computed tomography of pulmonary valve showing thickening of the 3 cusps (A) with commissural fusion (arrow) (B). Abbreviations as in Figures 2 and 6.

**FIGURE 8 Cardiac Computed Tomography**




Cardiac computed tomography showed right ventricular hypertrophy. Abbreviations as in Figures 4 and 5.

## CONCLUSIONS

Rheumatic heart disease is still a significant cause of valvular dysfunction in developing countries. It should be kept in the differential even if there is atypical presentation, especially in the presence of typical imaging features in patients from the endemic regions. Proper diagnosis and timely intervention can improve quality of life and prevent complications in patients with rheumatic heart disease.

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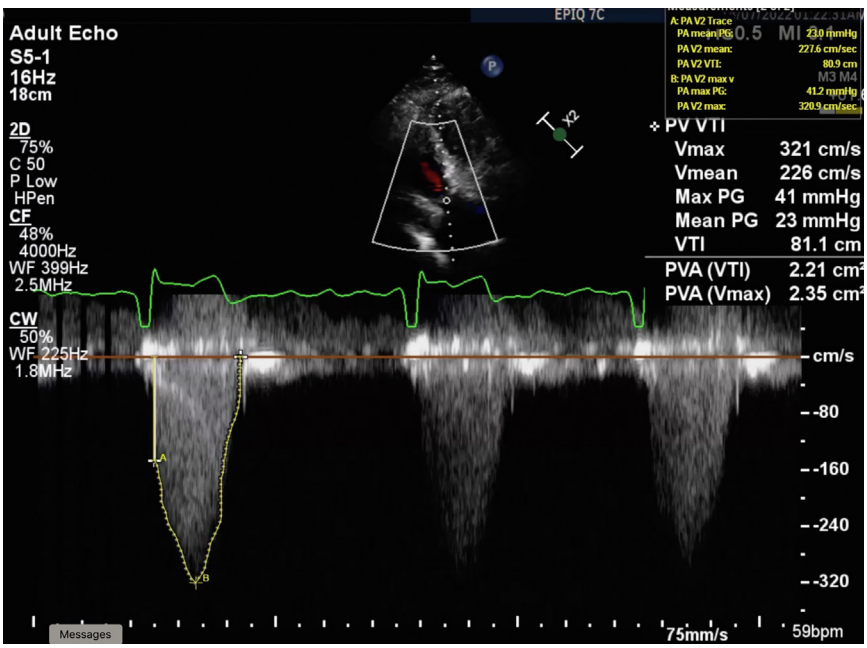
**FIGURE 9 Continuous Wave**




Continuous-wave Doppler postintervention showing the decrease in velocity and peak pressure gradient across the pulmonary valve.

## REFERENCES

**1.** Ruckdeschel E, Kim YY. Pulmonary valve stenosis in the adult patient: pathophysiology, diagnosis and management. *Heart*. 2019;105:414-422.

**2.** Muthiah R. Isolated rheumatic pulmonary valve disease—case reports. *Case Reports Clin Med*. 2016;5:207-215.

**3.** Buendía A, Attie F, Zabal C, Calderón-Colmenero J, Aranda A, Reyes P. Pulmonary valvar involvement in rheumatic heart disease—a study of 100 post-mortem cases. *Cardiol Young*. 1992;2:147-151.

**4.** Otto CM, Nishimura RA, Bonow RO, et al. 2020 ACC/AHA guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2021;77(4):e25-e197.

**5.** Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC guidelines for the management of adult congenital heart disease. *Eur Heart J*. 2020;42:563-645.

**KEY WORDS** **echocardiography, pulmonary valve, rheumatic heart disease**

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