

IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE

Rare Case of Coincident Quadricuspid Pulmonic Valve and Suprapulmonic Ridge



Multimodality Imaging

Abra Guo, MD,^a Hilmi Alnsasra, MD,^b Diego Medvedofsky, MD,^b Samer S. Najjar, MD,^b Monvadi B. Srichai, MD^{b,c}

ABSTRACT

A 32-year-old woman with a history of mild congenital pulmonic stenosis presented with new dyspnea on exertion. Multimodal imaging revealed a quadricuspid pulmonic valve with supralvalvular ridge. We illustrate the evaluation and diagnosis of this rare condition. (**Level of Difficulty: Advanced.**) (*J Am Coll Cardiol Case Rep* 2020;2:2196-8)
© 2020 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/>).

A 32-year-old woman with a history of mild congenital pulmonic stenosis and orthopnea of 6 years duration presented with 4 months of dyspnea on exertion. Recent chest computed tomography showed a dilated pulmonary artery measuring 3.7 cm. Physical exam was significant for a 2/6 systolic murmur but was otherwise benign. Her medical history was only significant for mild asthma.

The patient initially underwent transthoracic echocardiography (TTE), which showed a normal left ventricle, with ejection fraction of 60% to 65%. Mild thickening of the pulmonic valve was also noted, with mild stenosis (increased peak velocity of 2.4 m/s and mean gradient of 13 mm Hg). However, in some views of the TTE, there appeared to be a supralvalvular structure that was felt to represent supralvalvular stenosis instead of valvular stenosis (**Figure 1A, Video 1**). Flow acceleration was also noted through the pulmonic valve on color Doppler (**Figure 1B, Video 2**). Subsequent cardiac magnetic resonance revealed quadricuspid pulmonic valve (QPV) with mild-to-moderate pulmonic regurgitation as well as a supralvalvular ridge 1.3 cm above the pulmonic valve annulus (**Figures 1C and 1D, Videos 3 and 4**). This was associated with mild flow acceleration (peak and mean gradients of 21 mm Hg and 8 mm Hg, respectively) and dilation of the main pulmonary artery (diameter of 3.7 cm). No congenital shunt lesion was seen.

Transesophageal echocardiography was done to assess for right-to-left shunting, which was absent (**Figure 1E, Video 5**). Computed tomography angiography revealed normal coronary arteries, moderately dilated right ventricle with flattened interventricular septum, and QPV (**Figure 1F**). Right ventricular angiography demonstrated mild doming of the pulmonary valve leaflets as well as thin supralvalvular raphe attaching the leaflets to the sinotubular junction, consistent with mild valvular dysplasia (**Video 6**). The patient's work-up did not identify any cardiac etiology of her new dyspnea.

From the ^aDepartment of Internal Medicine, Medstar Georgetown University Hospital, Washington, DC; ^bMedStar Heart and Vascular Institute, MedStar Washington Hospital Center, Washington, DC; and the ^cDepartment of Cardiology, Medstar Georgetown University Hospital, Washington, DC.

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 *JACC: Case Reports* [author instructions page](#).

Manuscript received September 10, 2020; accepted September 22, 2020.

QPV is a rare cardiac finding that is often identified postmortem, as it is typically clinically asymptomatic. Congenital QPV is seen in 0.021% of the population, with a 2:1 incidence in men versus women (1). Anatomically, there is variation seen in QPV, the most common variant being 3 equal-size cusps with a smaller fourth cusp (60%) (1). Other cases have reported 4 (12%), and 2 larger cusps with 2 smaller cusps (15%) (1). Our patient was found to have 4 equally sized cusps. QPV can be seen in isolation or with other congenital heart conditions including atrial septal defect, ventricular septal defect, and patent ductus arteriosus. It can also be seen with pulmonic stenosis or regurgitation (3). Only about 4% of QPVs are associated with severe valvular dysfunction (2).

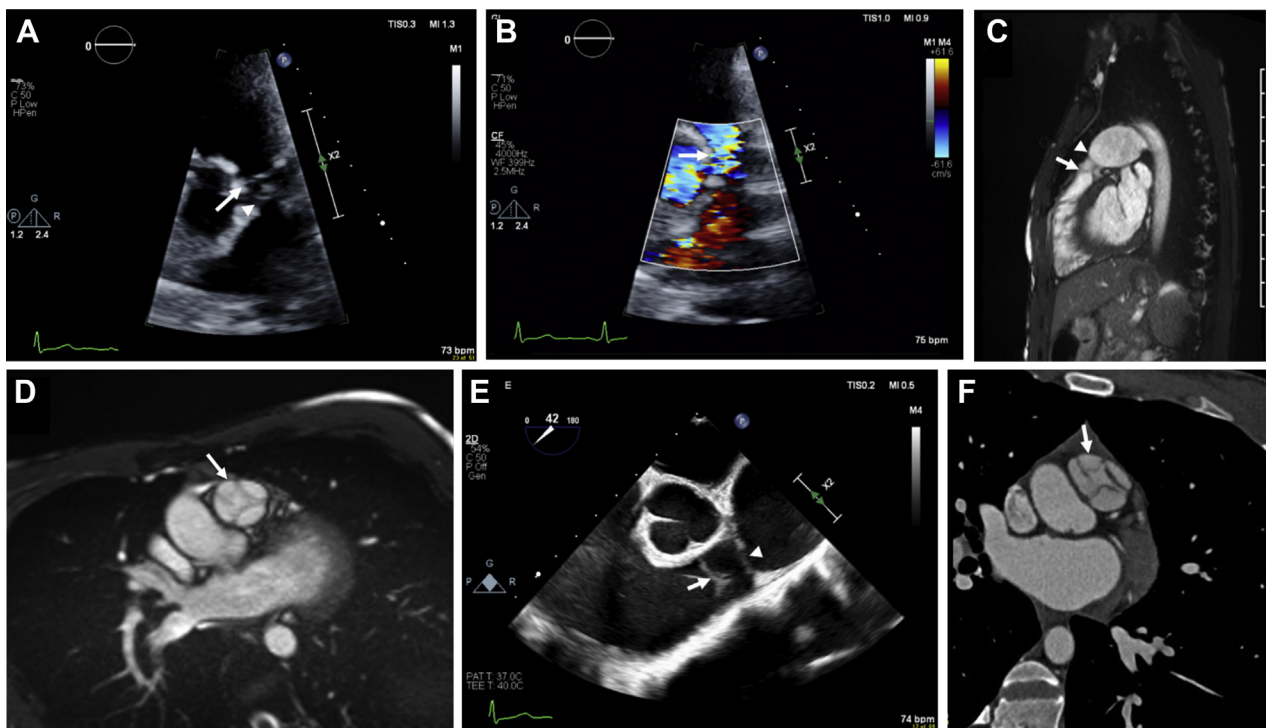
QPV is often underdiagnosed on TTE, given the anatomic relation between the pulmonic valve and the echocardiogram window (1). Transesophageal echocardiography is used to maximize view of the pulmonic valve. This can be achieved in the short-axis view by anteroflexing the probe between 135° and 145° (1). Cardiac magnetic resonance and computed tomography angiography are also helpful diagnostic modalities, particularly given their unrestricted imaging planes and field of view.

No formal recommendations exist for follow-up and management of QPV. To the best of our knowledge, this is the first case of QPV seen in conjunction with a supravulvar ridge. We plan to follow this patient with repeat echocardiography in 3 to 5 years. If any major changes are found, we may pursue more advanced imaging such as computed tomography or cardiac magnetic resonance.

**ABBREVIATIONS
 AND ACRONYMS**

QPV = quadricuspid pulmonic valve
TTE = transthoracic echocardiography

FIGURE 1 Multimodal Imaging of Quadricuspid Pulmonic Valve and Supravulvar Ridge



(A) Transthoracic echocardiography parasternal short-axis view. **Arrow** refers to pulmonic valve. **Arrowhead** refers to supravulvar ridge. **(B)** Transthoracic echocardiography parasternal short-axis view with color Doppler. **Arrow** refers to flow acceleration at the level of the pulmonic valve and supravulvar ridge. **(C)** Cardiac magnetic resonance oblique sagittal view demonstrating main pulmonary artery in long axis. **Arrow** refers to pulmonic valve. **Arrowhead** refers to supravulvar ridge. **(D)** Cardiac magnetic resonance oblique axial view demonstrating the pulmonic valve in short axis. **Arrow** refers to quadricuspid pulmonic valve. **(E)** Transesophageal echocardiography at 42°. **Arrow** refers to pulmonic valve. **Arrowhead** refers to supravulvar ridge. **(F)** Computed tomography angiography. **Arrow** refers to quadricuspid pulmonic valve.

AUTHOR DISCLOSURES


The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr. Barbara Srichai, Pasquerilla Healthcare Center, 5th Floor, 3800 Reservoir Road NW, Washington, DC 20007. E-mail: barbara.srichai@medstar.net.

REFERENCES

1. Dunay SN, Roberge RA, Avedissian LS. Quadricuspid pulmonic valve found on well exam. *Mil Med Res* 2015;2:10.
2. Manuel AM, Ribeiro J, Braga P, Fontes-Carvalho R. Quadricuspid pulmonary valve in a 50-year-old female with dyspnoea: innocent bystander or active player in disease? *Eur Heart J* 2019;40:3689.
3. Wang SS, Xu MG, Zhuang J, et al. Transthoracic echocardiographic evaluation of pulmonary valve anomalies in pediatric patients. *J Ultrasound Med* 2019;38:1091-6.

KEY WORDS cardiac magnetic resonance, echocardiography, pulmonic valve

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