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 supravalvular structure that was felt to represent supravalvular 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 supravalvular 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 supravalvular 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.
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 supravalvular 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.
AUTHOR DISCLOSURES

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

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KEY WORDS cardiac magnetic resonance, echocardiography, pulmonic valve

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