A 17-year-old female presented with a history of New York Heart Association class III breathlessness. On examination, a grade 4/6 systolic murmur was present in the aortic area. On transthoracic echocardiography, the left ventricle was normal size with normal ejection fraction and concentric hypertrophy (Fig. 1). The aortic valve (AV) was unicuspid in morphology. A single commissure was present at the fusion area of the left and non-coronary cusps (Figs. 2 and 3, Supplementary Video 1 in the online-only Data Supplement).

Severe aortic stenosis (AS) was identified. On suprasternal view, using the AV velocity time integral, the maximum pressure gradient (PG) across the valve was 219 mm Hg, aortic valve maximum velocity was 7.4 m/s, and mean PG was 134 mm Hg (Fig. 4, Supplementary Videos 2 and 3 in the online-only Data Supplement). The aortic annulus and root were normal size. The ascending aorta (maximum diameter: 39 mm) and the arch of the aorta (maximum diameter: 39 mm) were dilated (Fig. 5).

Retrospective series of patients with unicuspid AV showed an estimated incidence of 0.02%, with the most common lesion being AS; this condition required surgical correction most commonly in the third decade of life [1,2].

Supplementary Video Legends

Video 1. Showing parasternal long axis view and short axis view at aortic valve level using biplane method to demonstrate valve morphology.

Video 2. Apical 5-chamber view with color Doppler showing turbulence across the aortic valve.

Video 3. Suprasternal view with color Doppler showing turbulence across the aortic valve.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.22468/cvia.2021.00136.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Acknowledgments

None.

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Fig. 1. Parasternal long-axis view demonstrating concentric LV hypertrophy. LA: left atrium, LV: left ventricle, Ao: aorta.

Fig. 2. Parasternal short-axis view showing a unicuspid aortic valve in systole. LA: left atrium, RA: right atrium, RV: right ventricle.

Fig. 3. Parasternal short-axis view showing a unicuspid aortic valve in diastole with a single commissure. LA: left atrium, RA: right atrium, RV: right ventricle.

Fig. 4. A continuous wave Doppler jet across the aortic valve in the suprasternal view. AV: aortic valve, Vmax: maximum velocity, PG: pressure gradient.

Fig. 5. Modified parasternal view showing a dilated ascending aorta.