

Rarer Than a Unicorn: 3D TEE Diagnosis a Unicuspid Aortic Valve with Associated Aortic Aneurysm

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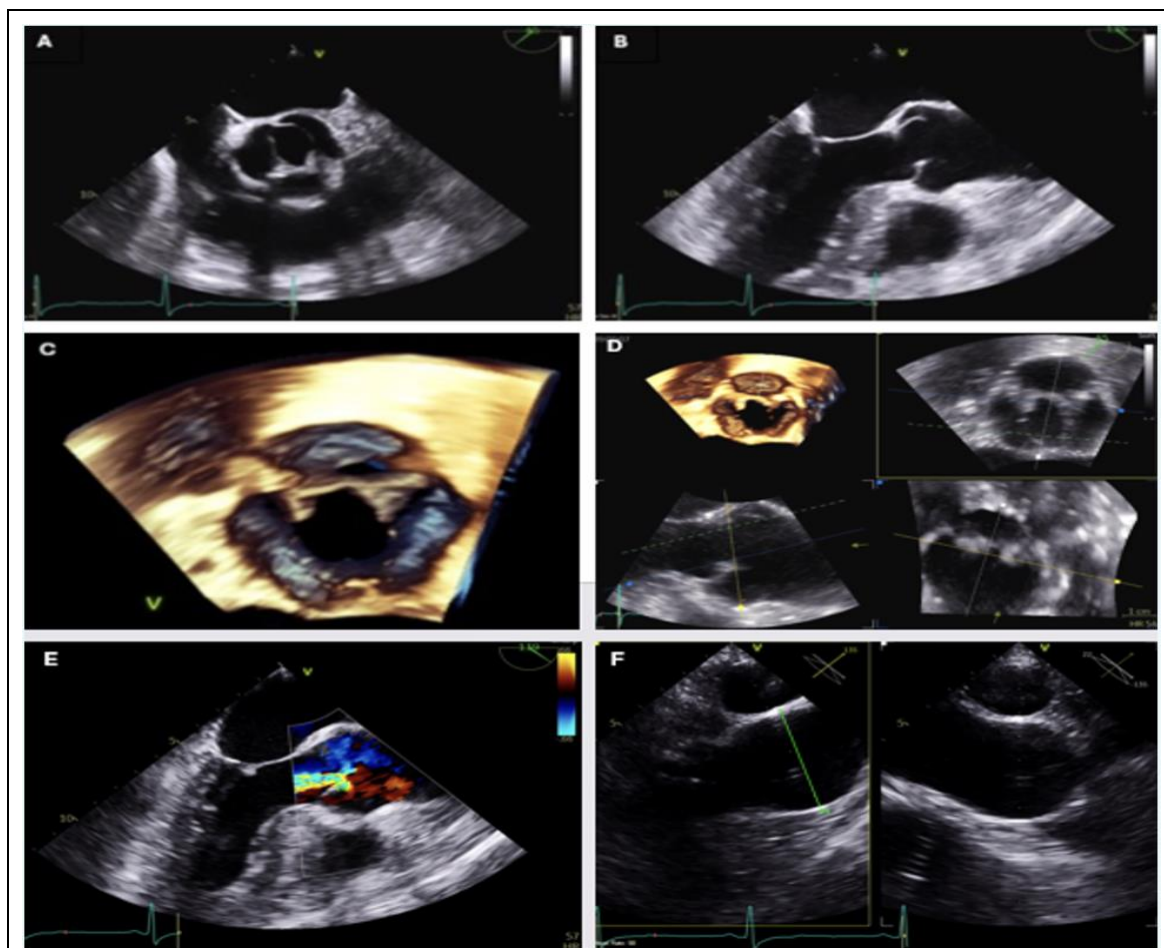


Figure 1: Panel A: Short-axis view showing sclerosis and cusp fusion. **Panel B:** Long-axis view illustrating leaflet systolic doming. **Panels C-D:** 3D multiplanar reconstruction confirming valve morphology and measuring area (1.8 cm²). **Panel E:** Color Doppler showing central regurgitant jet (vena contracta 0.47 cm). **Panel F:** Upper esophageal view showing ascending aortic aneurysm (4.3 cm).

Case Presentation

A 67-year-old man with hypertension, dyslipidemia, chronic kidney disease, and coronary calcifications presented with progressive exertional dyspnea in the setting of mild mixed aortic valve disease. Pharmacologic nuclear stress testing showed no ischemia. Transthoracic echocardiography (TTE) was limited but showed normal chamber size and function, moderate aortic regurgitation, and mild-to-moderate aortic stenosis.

Transesophageal echocardiography (TEE) revealed leaflet sclerosis, systolic doming, and commissural fusion between the noncoronary and right cusps, as well as the right and left cusps (Figure 1A-B; Videos 1-2). 2D and 3D multiplanar reconstruction confirmed a single-commissure unicuspid valve (Figure 1A-D; Video 3), with moderate central regurgitation (Figure 1E; Video 4) and mild stenosis (valve area 1.8 cm²). A proximal ascending aortic aneurysm was visualized, measuring 4.3 cm (Figure 1F). A CT scan is pending to further assess the aortic root aneurysm and valve morphology.

Keywords: Unicuspid aortic valve; Mixed aortic disease; Aortic aneurysm; 3D TEE

Discussion

Single commissure aortic valve is rare, with an incidence of 2 per 10,000 people, though presents in 1 in 20 surgically removed stenotic valves, indicating under-recognition [1,2]. Prompt diagnosis is important given the condition's association with congenital conditions such as aortopathy, coarctation, coronary anomalies and complications including dissection [3].

Similar to bicuspid valves, aortopathy in UAV is associated with intrinsic defects from disrupted elastin–collagen structure and increased matrix metalloproteinase activity as well as markedly abnormal flow hemodynamics and elevated wall shear stress (WSS) contributing to progressive aortic wall remodeling and dilatation. While 3D TEE can provide remarkable anatomical detail with reasonable spatial and temporal resolution, CT and 4D flow MRI allow comprehensive visualization of the thoracic aorta, quantification of flow dynamics, and WSS assessment, making them necessary for UAV-related aortopathy evaluation and surveillance [4]. Given the condition's rarity, no distinct surgical cutoffs currently exist for UAV associated valvulopathy and aortopathy beyond available recommendations for trileaflet and bicuspid valves.

Patient Consent: Written informed consent obtained.

Videos: (All associated clinical videos are available online with the published article).

Video 1: Short-axis TEE view.

Video 2: Long-axis TEE view.

Video 3: 3D/3D multiplanar reconstruction.

Video 4: Color-flow Doppler.

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