

Unicuspid Aortic Valve Stenosis in a Child

Rajeev Bhardwaj*

Received: 20 April 2025; Accepted: 09 August 2025



A 15-year-old male presented with a history of dyspnea for the last 1 year. Dyspnea progressed gradually, and he deteriorated to New York Heart Association

(NYHA) class III for the last 3 months. History of paroxysmal nocturnal dyspnea (PND) was present for the last 1 month. He also had a history of dizziness during playing.

His electrocardiogram (ECG) showed left ventricular hypertrophy with strain. His echocardiography showed the presence of unicuspid aortic valve (UAV) in parasternal short axis view, with typical teardrop appearance in systole (Fig. 1). The valve was unicommissural (Fig. 2). The peak gradient across the valve was 71 mm Hg, and the mean gradient was 46 mm Hg (Fig. 3). Left ventricular ejection fraction was 19.4%.

The aortic valve normally has three cusps. Bicuspid aortic valve is the most common anomaly found in the aortic valve. UAV is a form of anomaly that is very rare and is found only in around 0.02% patients with aortic stenosis when echocardiography is used to find the prevalence and around 5% of patients when surgical series are seen.¹ On the basis of number of commissures, it is of two forms: Unicuspid unicommissural (UUAV), when one commissure is present, and when no commissure is present, it is called unicuspid acommisural (UAAV).² However, since many patients are asymptomatic, the true incidence in population is not known and may be much more than seen on echocardiography or at surgery.³ UAAV form presents early in life, whereas UUAV has relatively late presentation. So, if seen in adults, it is almost always of UUAV variety.⁴ Collins et al.⁵ have shown that decreased number of cusps in the aortic valve cause more pathological changes in cusps as well as in aorta. Patients with UAV may present with pure aortic stenosis, pure aortic regurgitation, but more commonly with mixed lesion (aortic stenosis with aortic regurgitation).⁶ Our patient had pure aortic stenosis and was of UUAV type.

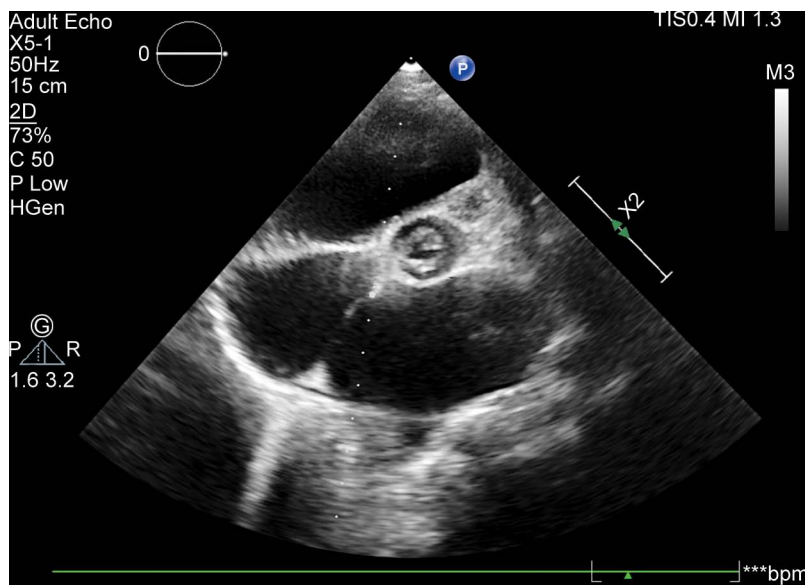


Fig. 1: Parasternal short axis view showing UAV in systole

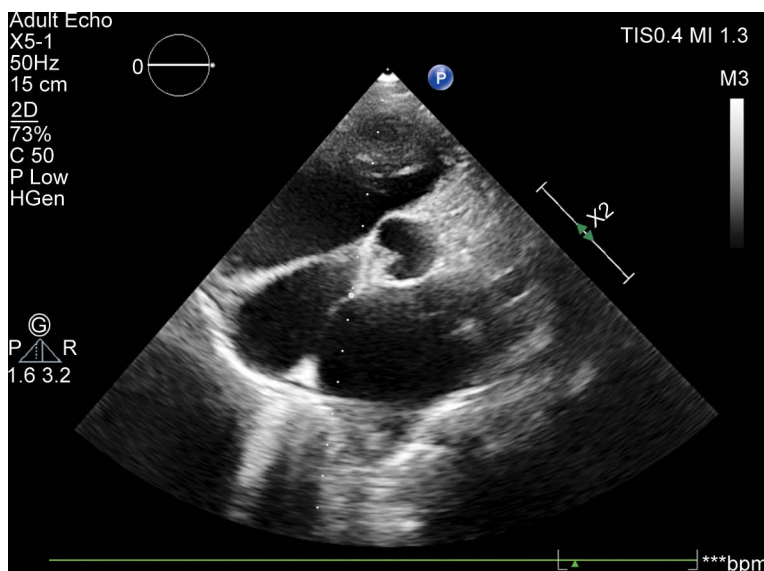


Fig. 2: Parasternal short axis view in diastole showing unicommissural unicuspid valve

Professor and Head of Cardiology, Department of Cardiology, MM Institute of Medical Sciences & Research, Ambala, Haryana, India; *Corresponding Author

How to cite this article: Bhardwaj R. Unicuspid Aortic Valve Stenosis in a Child. J Assoc Physicians India 2025;73(11):89–90.

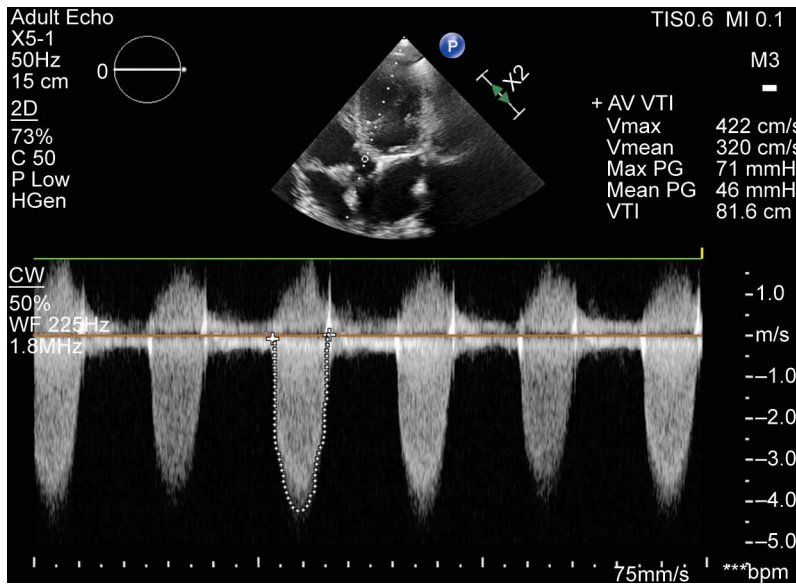


Fig. 3: Continuous wave Doppler showing gradients across the valve, suggesting severe aortic stenosis

REFERENCES

1. Novaro GM, Mishra M, Griffin BP. Incidence and echocardiographic features of congenital unicuspid aortic valve in an adult population. *J Heart Valve Dis* 2003;12(6):674–678.
2. Anderson RH. Understanding the structure of unicuspid and unicommissural aortic valve. *J Heart Valve Dis* 2003;12(6):670–673.
3. Novaro GM, Mishra M, Griffin BP. Incidence and echocardiographic features of congenital unicuspid aortic valve in an adult population. *J Heart Valve Dis* 2003;12:674–678.
4. Matsumoto K, Tanaka H, Hiraishi M, et al. A case of unicommissural unicuspid aortic valve stenosis diagnosed by real time three-dimensional transesophageal echocardiography. *Echocardiography* 2011;28(8):E172–E173.
5. Collins MJ, Butany J, Borger MA, et al. Implications of a congenitally abnormal valve: a study of 1025 consecutively excised aortic valves. *J Clin Pathol* 2008;61:530–536.
6. Pan J. Unicuspid aortic valve: a rare congenital anomaly. *Cardiology* 2022;147(2):207–215.