

Unicuspid Aortic Valve in a Young Woman Associated with Aortopathy and Atrial Septal Defect: Therapeutic Implications

Karen Queiroz dos Santos,¹  Marcelo Barbosa Luckemeyer de Melo¹ 

Rede Saúde Mais,¹ Alto Araguaia, MT – Brazil

Abstract

Unicuspid aortic valve (UAV) is a rare congenital anomaly, frequently associated with early progression to severe aortic stenosis and aortopathy. We report the case of a 38-year-old woman who presented with difficult-to-control hypertension and progressive exertional dyspnea. Transthoracic echocardiography revealed critical aortic stenosis (peak gradient of 183 mmHg, mean gradient of 125 mmHg, and valve area of 0.59 cm²), associated with mild aortic regurgitation and ascending aortic dilation (45–46 mm). Three-dimensional transesophageal echocardiography confirmed a unicommissural UAV, as well as an ostium secundum atrial septal defect with a left-to-right shunt (Qp/Qs = 2.3). Given the significant hemodynamic impact, aortic valve replacement with a bioprosthetic valve was performed. The patient developed a conduction disturbance during the postoperative period, requiring permanent pacemaker implantation. At the 6-month follow-up, she was asymptomatic, and the prosthetic valve was functioning normally. This case illustrates the importance of advanced echocardiography in identifying rare valvular anomalies and highlights the uncommon association between UAV, aortopathy, and atrial septal defect in a young woman, with relevant implications for clinical and surgical management.

Introduction

Unicuspid aortic valve (UAV) is a rare congenital malformation, with an estimated prevalence of less than 0.02% in the general population.^{1,2} It is characterized by a more aggressive clinical course than that associated with bicuspid aortic valve, with early degeneration and rapid progression to severe aortic stenosis in young individuals.^{2,3} UAV is frequently associated with ascending aortic dilation and, more rarely, with other congenital heart defects, posing a diagnostic challenge. Transesophageal echocardiography, especially with three-dimensional reconstruction, plays a fundamental role in identifying this anomaly and differentiating it from a calcified bicuspid valve.^{4,5} The association of UAV, aortopathy, and other congenital

anomalies in a young woman is uncommon, underscoring the clinical relevance of this case report.

Case report

We report the case of a 38-year-old woman with difficult-to-control hypertension who presented with progressive dyspnea upon moderate exertion. As shown in Figure 1, transthoracic echocardiography revealed critical aortic stenosis (peak gradient of 183 mmHg, mean gradient of 125 mmHg, and valve area of 0.59 cm²), associated with mild aortic regurgitation and ascending aortic dilation (46 mm). Concentric left ventricular hypertrophy, preserved ejection fraction, grade I diastolic dysfunction, reduced global longitudinal strain (13%), mild left atrial dilation, and signs of right heart volume overload were also observed. Another echocardiogram confirmed elevated transvalvular velocities and a significant reduction in valve area (Figure 2). Three-dimensional transesophageal echocardiography confirmed a unicommissural UAV (Figure 3) and an ostium secundum atrial septal defect (14 mm), with a left-to-right shunt and a Qp/Qs ratio of 2.3, indicating a significant hemodynamic impact. Given this clinical condition, aortic valve replacement was indicated. Following shared decision-making, taking the patient's age, preferences, and desire to avoid chronic anticoagulation into account, a bioprosthetic valve was selected, which is considered an acceptable strategy under current guidelines.^{1,4} Although the ascending aorta was dilated, its diameter was within the borderline range for isolated surgical intervention; therefore, clinical follow-up and serial imaging were chosen. During the postoperative period, she developed a conduction disturbance, requiring permanent pacemaker implantation. At the 6-month outpatient follow-up, she was asymptomatic, with a normally functioning prosthetic valve.

Discussion

UAV is an extremely rare congenital malformation, often associated with a more aggressive clinical course than bicuspid aortic valve, and requiring early intervention.^{2,3} Consistent with the findings described by Roberts et al.,² presentation in young adulthood with critical aortic stenosis underscores the accelerated nature of valvular degeneration in these patients. From an anatomical perspective, the unicommissural type is the most commonly seen in patients who survive into adulthood.³ Identifying this anomaly via transthoracic echocardiography can be challenging; transesophageal echocardiography, particularly with three-dimensional reconstruction, is crucial for differential diagnosis, in accordance with current guidelines.^{4,5} The association between UAV and ascending aortic dilation suggests an underlying intrinsic aortopathy, possibly related to structural alterations in the extracellular matrix of the vascular wall. This mechanism is similar to that observed in bicuspid aortic valve.^{6,7}

Keywords

Aortic Valve; Aortic Valve Stenosis; Aortic Aneurysm; Congenital Heart Defects

Mailing Address: Karen Queiroz dos Santos •

Rede Saúde Mais. Avenida Vereadora Maria da Glória Favero, nº 586. Postal code: 78780-000. Bairro Vila Aeroporto, Alto Araguaia, MT – Brazil

E-mail: karenqueirozsantos@gmail.com

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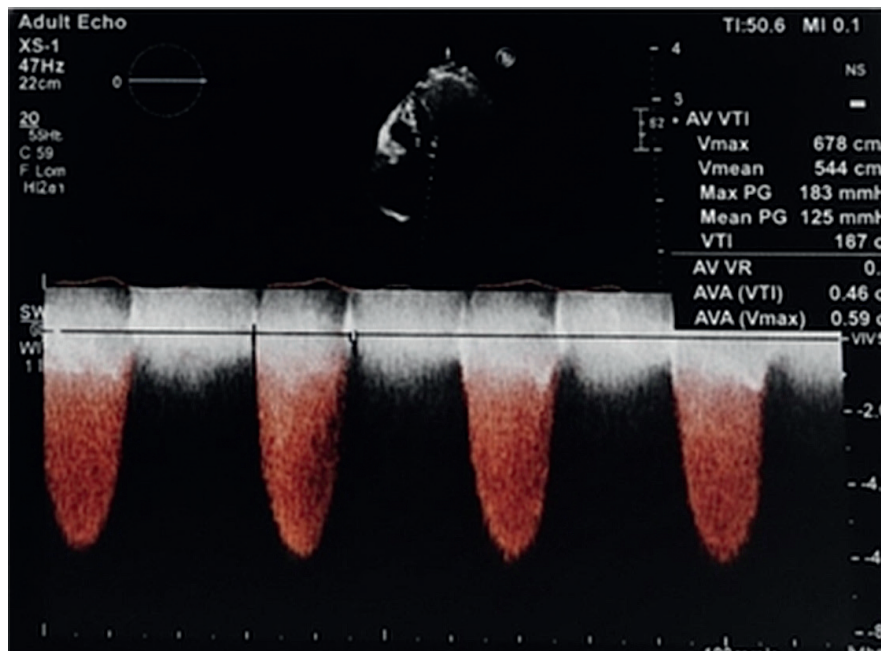


Figure 1 – Transthoracic echocardiography with continuous-wave Doppler (apical five-chamber view) demonstrating transvalvular aortic flow with significantly elevated velocities, consistent with critical aortic stenosis. Peak systolic gradient: 183 mmHg; mean gradient: 125 mmHg; aortic valve area: 0.59 cm². AV VTI: Aortic Valve Velocity-Time Integral; AV: Aortic Valve; VR: Velocity Ratio; AVA: Aortic Valve Area.

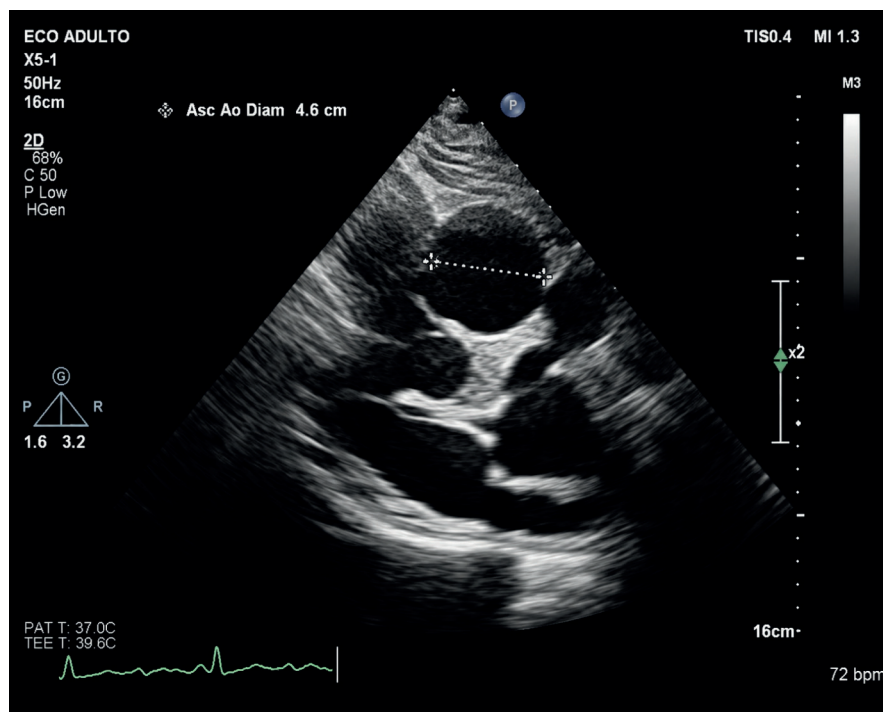


Figure 2 – Transthoracic echocardiogram showing ascending aortic dilation (arrow), with a maximum diameter of 46 mm, consistent with aortopathy associated with a UAV. PAT: Patient Temperature; TEE: Transesophageal Echocardiography.

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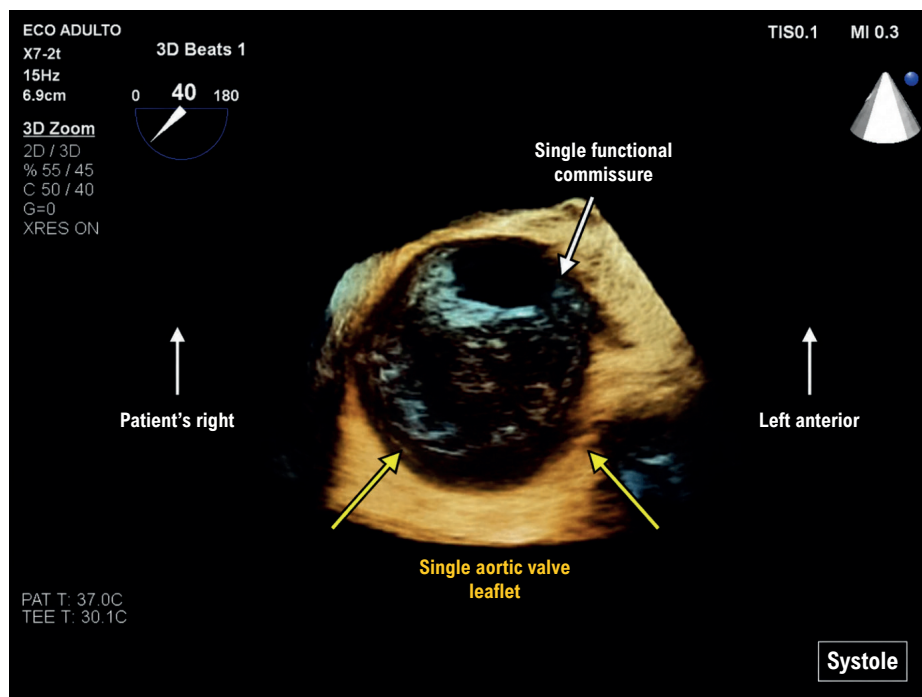


Figure 3 – Three-dimensional transesophageal echocardiography (en face view) of the aortic valve, acquired during systole, showing a unicommissural UAV. The white arrow indicates a single functional commissure, allowing for differentiation from a calcified bicuspid aortic valve. PAT: Patient Temperature; TEE: Transesophageal Echocardiography.

Furthermore, reduced global longitudinal strain (13%),⁸ even with a preserved ejection fraction, indicates subclinical myocardial dysfunction, as described by Delgado et al.,⁹ representing an early marker of ventricular impairment and greater hemodynamic severity. The association of UAV, aortopathy, and atrial septal defect is uncommon in clinical practice, particularly in patients who reach adulthood without a prior diagnosis. This finding suggests a broader spectrum of cardiac developmental abnormalities. The severity of aortic stenosis warrants a surgical approach, as recommended by current guidelines.^{1,4,10} The choice of a bioprosthetic valve, although less durable, is acceptable given the patient's preference and the risks associated with chronic anticoagulation.^{1,4} Postoperative complications, including conduction disturbances, have been documented in the literature and should be taken into account during preoperative management and counseling. The reported case reinforces the importance of diagnostic suspicion in young patients with significant aortic stenosis, as well as the key role of advanced echocardiography in anatomical definition, prognostic stratification, and therapeutic planning.

Author Contributions

Conception and design of the research and writing of the manuscript: Santos KQ; acquisition of data, analysis and interpretation of the data and critical revision of the manuscript for intellectual content: Santos KQ, Melo MBL.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

The authors declares that this clinical case report does not constitute research involving human subjects. Informed consent was obtained from all participants included in the study.

Use of Artificial Intelligence

The authors did not use any artificial intelligence tools in the development of this work.

Availability of Research Data

The underlying content of the research text is contained within the manuscript.

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