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Severe stenosis of a unicuspid aortic valve

Short title: Stenosis of unicuspid aortic valve

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We present a case of a 42-year-old female with a unicuspid aortic valve identified by echocardiography and confirmed by cardiac computed tomography. This woman was referred to our Clinic for diagnostic workup of progressive significant breathlessness at exercise and suspected arrhythmia. On admission, she was in stable clinical condition without significant abnormalities on physical examination except for an loud systolic murmur at the aortic valve.

A transthoracic echocardiography (2D TTE) revealed an unicuspid, unicommissural aortic valve with severe, high-gradient aortic stenosis. The aortic valve area (AVA) was 0.9 cm² with a peak velocity of 4.4 m/s with a mean gradient of 42 mmHg. Additional findings of mild aortic regurgitation and dilatation of the ascending aorta (51 mm) were found with no signs of aortic

coarctation. A transesophageal echocardiography (2D, 3D TEE) confirmed the defect in the aortic valve. Findings were consistent for severe aortic stenosis. For surgical planning, the patient underwent a computed tomography (CT) which showed no evidence of coronary artery stenosis and confirmed unicuspid aortic valve.

During cardiac surgery replacement of the unicuspid aortic valve was performed and biological prosthesis was implanted. Moreover ascending aorta was replaced with graft.

Unicuspid, unicommissural aortic valve is an extremely rare congenital anomaly of the aortic valve [1]. The estimated frequency of its occurrence is 0.02% and is 100 times less common than a bicuspid aortic valve (BAV). Importantly, unicuspid aortic valves are associated with rapid progression of valvular dysfunction and aortic dilatation.

In the case of our patient, the performed examinations (TTE, TEE CT) showed the three major criteria of a unicuspid valve: single commissural zone of attachment, rounded leaflet-free edge on the opposite side of the commissural attachment zone, and eccentric valvular orifice during systole [2].

In conclusion, this case report confirms the steps to accurately assess a rare defect like unicuspid aortic valves using accessible non-invasive methods.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

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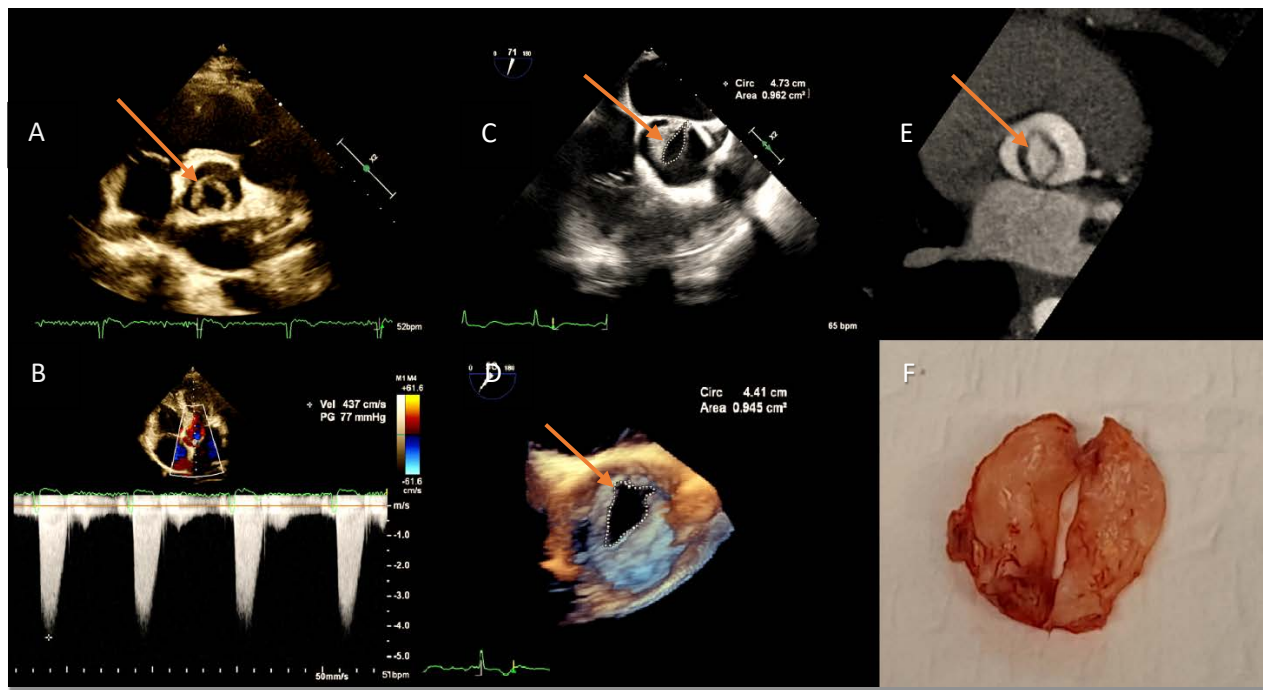


Figure 1. **A.** Unicuspid aortic valve (the arrow); short axis view, 2D TTE. **B.** A peak velocity >4 m/sec measured using continuous wave Doppler, TTE. **C.** Measurement of an aortic-valve area using planimetric two-dimensional (2D) TOE. **D.** Measurement of an aortic-valve area using planimetric three-dimensional (3D) TOE. **E.** CT image of unicuspid aortic valve. **F.** Unicuspid aortic valve after cardiac surgery

Abbreviations: 2D TTE, two dimensional transthoracic echocardiography; 3D-TOE- three dimensional transoesophageal echocardiography; CT, computed tomography