

# Unicuspid aortic valve and infective endocarditis

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## Abstract

Abstract Unicuspid aortic valve (UAV) is a rare congenital malformation, related to an aggressive evolution of valve function. Its diagnosis represents an important cardiac imaging challenge, since it is often difficult to differentiate between other anomalies like bicuspid aortic valve. Recently, bicuspid aortic valve has been associated with IE, especially when aortic valve function shows significant stenosis, however, the relationship between IE and UAV is unclear. The anatomical characteristics of UAV could predispose to IE. This report describes a young patient affected by infective endocarditis (IE) with UAV and the case reviews the echocardiographic criteria used to improve its management.

## TITLE PAGE

Article Title:

UNICUSPID AORTIC VALVE AND INFECTIVE ENDOCARDITIS

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## Abstract

Unicuspid aortic valve (UAV) is a rare congenital malformation, related to an aggressive evolution of valve function. Its diagnosis represents an important cardiac imaging challenge, since it is often difficult to differentiate between other anomalies like bicuspid aortic valve. Recently, bicuspid aortic valve has been associated with IE, especially when aortic valve function shows significant stenosis, however, the relationship between

IE and UAV is unclear. The anatomical characteristics of UAV could predispose to IE. This report describes a young patient affected by infective endocarditis (IE) with UAV and the case reviews the echocardiographic criteria used to improve its management.

## Keywords

Infective endocarditis

Aortic valve

Echocardiography

## Abbreviations

BAV: Bicuspid aortic valve

IE: Infective endocarditis

TEE: Transesophageal echocardiography

TTE: Transthoracic echocardiography

UAV: Unicuspid aortic valve

## Introduction

Unicuspid aortic valve (UAV) is an infrequent congenital anomaly secondary to altered aortic cusp development from tubercles<sup>1</sup>. A disordered number of cusps results from fusion or duplication, with a reported occurrence with echocardiography of 1% for bicuspid aortic valve (BAV), 0.05% for quadricuspid aortic valve, and 0.02% for UAV<sup>2</sup>. Due to its rarity, clinical progression and complications of UAV are not well known.

## Case Report

A 34-year-old woman with a two-week history of fever presented to the emergency department because of sudden-onset mild right hemiparesis and dysarthria. Computed tomography scan demonstrated a left thalamic stroke. Her symptoms rapidly resolved, thus intravenous thrombolysis was not prescribed. Blood tests revealed leukocytosis and elevated C-reactive protein levels. Cardiac systolic murmur was patent, and transthoracic echocardiography (TTE) showed an UAV with moderate aortic stenosis without regurgitation or aortic dilation. An irregular vibrated mass was also identified and transesophageal echocardiography (TEE) confirmed the presence of 6x8-mm vegetation attached to the aortic valve (Figure 1A; arrows). Blood cultures showed *Staphylococcus epidermidis*, supporting a diagnosis of infective endocarditis (IE). After two weeks of medical treatment, aortic valve replacement was indicated for persistent vegetation after an embolic episode. Direct surgical inspection of the aortic valve confirmed a stenotic UAV. No postoperative complications occurred, and the patient was discharged after finishing the antibiotic therapy.

## Discussion

UAV is a rare congenital malformation, resulted from the abnormal fusion of the three tubercles. The true prevalence of UAV is unknown and likely underestimated, since it is often difficult to differentiate between other anomalies like BAV by cardiac imaging<sup>3,4</sup>. Further, >50% of UAV cases are only confirmed by intraoperative surgery or autopsy<sup>5</sup>. Two phenotypes of UAV have been described in the literature, the unicommissural type with a single leaflet from the aortic wall and one being the pinhole-shaped aocommissural UAV<sup>6,7</sup>. Four echocardiographic criteria are used to improve the differential diagnosis<sup>8</sup>: (i) single commissural attachment zone, (ii) rounded leaflet-free edge on the opposite side of the commissural attachment zone, (iii) eccentric valvular orifice during systole and (iv) age <20 years and mean transvalvular gradient >15mmHg. When three of them are met, they have 97% specificity for diagnosing of UAV (Figure 1B; Videos 1-4). Clinically, UAV presents a more aggressive evolution during follow-up than BAV—the valve is rarely normofunctional and generates earlier significant aortic stenosis and more aortic valve intervention associated with ascending aortic aneurysm repair via the Bentall procedure than in BAV patients<sup>5,9</sup>.

Recently, BAV has been associated with IE, especially when aortic valve function shows significant stenosis<sup>10</sup>. This new evidence challenges the recommendations of antibiotic prophylaxis in BAV patients undergoing dental procedures. The high-risk profile of UAV for developing aortic complications highlight the differential diagnosis between both entities and the potential risk of IE should raise the question if antibiotic prophylaxis may be also recommended in these patients.

### Declaration of interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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### Ethical Approval

We anonymize all images and data.

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### FIGURE LEGENDS

**Figure 1A. Transesophageal echocardiography assessment of unicuspid aortic valve.** **Upper left**, Mid-esophageal 2D short-axis transesophageal echocardiography view of the aortic valve. LA, Left atrium; PV, Pulmonic valve; TV, Tricuspid valve. **Upper right**, Mid-esophageal 3D short-axis transesophageal echocardiography view of the aortic valve. LA, Left atrium; PV, Pulmonic valve; TV, Tricuspid valve. **Lower left**, Mid-esophageal 2D long-axis transesophageal echocardiography view of the aortic valve, showing the

vegetation (yellow arrow). AA, Ascending aorta; LVOT, Left ventricular outflow tract. **Lower right**, Mid-esophageal 3D long-axis transesophageal echocardiography view of the aortic valve, showing the vegetation (yellow arrow). AA, Ascending aorta.

**Figure 1B. Schematic of echocardiographic features to detect unicuspid aortic valve morphology.** Four criteria can be used to detect unicuspid aortic valve: (I) mean aortic transvalvular gradient  $>15$  mmHg and patient age of  $<20$  years, (II) single commissural zone of attachment, (III) eccentric valvular orifice during systole, and (IV) rounded leaflet-free edge on opposite side of the commissural attachment zone.

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