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One-leaf clover: a rare case of a unicuspid unicommissural aortic valve

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Congenital malformations of the aortic valve may present as bicuspid, quadricuspid, or unicuspid aortic valve (UAV) anomalies.[1] A UAV is an extremely rare type of malformation with an estimated annual prevalence of 0.02%.[2] It is often seen in adulthood, especially during the third to fifth decades of life, with presenting symptoms of severe aortic stenosis or aortic regurgitation.[1,3] We report a case of a 30-year-old male who presented with generalized fatigue, worsening dyspnoea on exertion, and palpitations who was found to have a unicuspid unicommissural aortic valve.

A 30-year-old male presented with generalized fatigue, palpitations, worsening dyspnoea on exertion, and syncope. He was found to have catecholaminergic polymorphic ventricular tachycardia with plans for implantable defibrillator placement. A transthoracic echocardiogram (TTE) revealed mild aortic stenosis and moderate aortic regurgitation with a poorly visualized aortic valve with concern for congenital aortic valve pathology. A transoesophageal echocardiogram (TOE) was done subsequently which revealed a UAV with a single commissure and two false commissures along with restricted

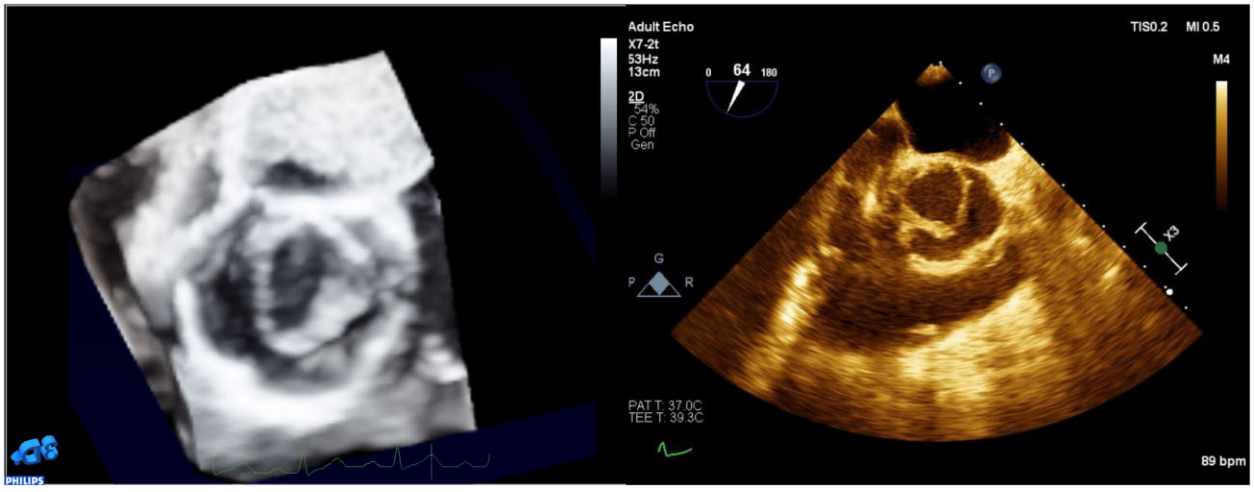

Figure 1 Left: A three-dimensional transoesophageal echocardiogram image of a unicommissural, unicuspid aortic valve in systole that is an en-face view from the aortic aspect. Right: A two-dimensional transoesophageal echocardiogram image in short axis (64 degrees). As the aortic valve opens, the eccentric opening reveals the absence of cusp separation. There is a single commissure of the aortic valve with two raphae (or false commissures) seen.

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Video 1 A three-dimensional transoesophageal echocardiogram video of a unicommissural, unicuspid aortic valve in systole that is an enface view from the aortic aspect.

aortic leaflet opening suggestive of mild aortic stenosis and moderate aortic regurgitation (*Figure 1*, *Video 1*).

Our imaging case highlights the rare TOE findings of a UAV. Unicuspid aortic valves can be divided into two distinct subtypes: acommissural and unicommissural.[2] Subtypes of UAV differ structurally and in time of symptomatic onset. An acommissural UAV is defined by a single leaflet, no commissure, and small central opening.[3] The onset of symptoms is during infancy and requires early intervention for survival. A unicommissural UAV is defined by a single commissure and one cusp. Diagnosis of a UAV can be done with two-dimensional (2D) or three-dimensional (3D) TTE or TOE, cardiac computed tomography, or cardiac magnetic resonance imaging.[1] The 3D TOE is the superior modality for diagnosis as UAVs typically present with severe calcification and obscure morphology that is difficult to differentiate with 2D TTE. Unicuspid aortic valves should be imaged during systole to capture the absence of cusp separation during aortic valve opening.[1]

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