Bicuspid aortic valve with vertical opening and double raphe

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A 17-year-old male patient presented with paroxysmal supraventricular tachycardia. Transthoracic echocardiography showed an abnormal aortic valve with mild aortic insufficiency. Transoesophageal echocardiography confirmed a functional bicuspid aortic valve however with a vertical opening and a double raphe (Figure 1). There were no signs of aortic aneurysm.

Normal bicuspid valves have a horizontal opening plane and one raphe as a result of one conjoint cusp. Bicuspid aortic valve disease is a congenital cardiac defect with a prevalence of 0.5–1% in a 2:1 male:female ratio. The echocardiographic diagnosis of a bicuspid aortic valve is based on the demonstration of two cusps and two commissures with a single coaptation line in a short-axis view. In the majority of cases, one cusp is positioned anteriorly and the second posteriorly with a horizontal coaptation line, a single raphe being usually located in the anterior cusp. Alternatively, the cusps are positioned to the right and left with a vertical, anterior–posterior coaptation line, a single raphe usually being located in the right cusp. The existence of a double raphe in a bicuspid valve with vertical opening is extremely rare in the continuum of dysmorphic aortic valves and has not been described before. It might be speculated that the valve defect was in congenital development a quadricuspid aortic valve which became a functional bicuspid valve after two cusps conjoint (Figure 1). The most frequent associated finding of the bicuspid valve is dilation of the proximal ascending aorta secondary to abnormalities of the aortic media. Complications include aortic valve stenosis or incompetence, endocarditis, aortic aneurysm formation, and aortic dissection. Prior studies have demonstrated that regular follow-up in adults with bicuspid aortic valves is essential to detect valve-related complications or aneurysm.

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

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