A rare anatomical form of quadricuspid aortic valve diagnosed by transthoracic echocardiography

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Dear Editor,

Quadricuspid aortic valve (QAV) is a rare congenital anomaly, far less common than bicuspid or unicuspid aortic valves. In a series of autopsies it ocurred in about 0.008% and 0,033% of cases (1). The first case was reported by Balington (2) in 1862. Since then only a few cases (3, 4) have been reported. Most of the cases are discovered incidentally at the time of aortography, aortic valve replacement surgery, or autopsy. A 22-year man was examined for symptoms of atypical angina in emergency service. He had no previous history of cardiovascular disease or drug use. Physical examination is normal. His blood pressure was 120/60 mmHg and pulse rate was 86 beats/min. On auscultation of the heart, there was a grade 1-2/4 diastolic murmur in aortic area. The electrocardiogram showed normal sinus rhythm and the chest radiogram was normal. Transthoracic echocardiography showed a QAV with three equal size cusps and one smaller cusp in the short-axis view (Fig. A). The left ventricular size and ejection fraction were normal and an eccentric mild aortic regurgitation in the long axis view (Fig. B). Our case is type B according to classification of QAV by Hurwitz and Roberts in 1973 (5). Sometimes diagnosis may be missed with trans-thoracic echocardiography and transesophageal approach is needed. Especially in young patients with aortic regurgitation, when the number of valve leaflets is not properly identified by TTE, a transesophageal echocardiogram should be performed. Management of patients with QAV is represented by strict follow-up, because those patients may require aortic valve replacement in their future life. QAV diagnosis before aortic valve replacement is important to guide surgical technique, being often associated to anomalous positioning of coronary ostia (6). There have been reports of endocarditis in patients with QAV, and patients with unequal cusps are considered at higher risk for this complication.

Key Words: Quadricuspid aortic valve, Transthoracic Echocardiography

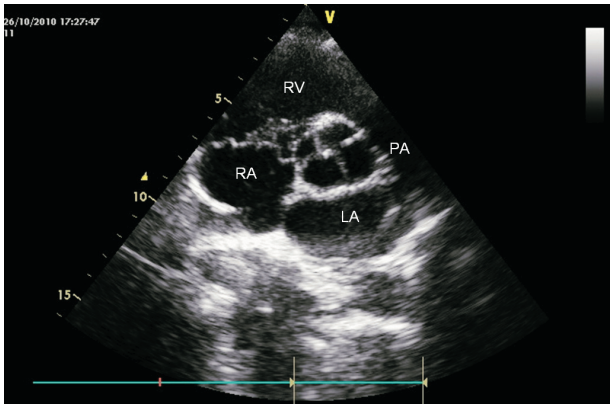

Fig. A Transthoracic echocardiography showed a QAV with three equal size cusps and one smaller cusp in the short-axis view . RV: Right Venticul, RA: Right Atrium, LA: Left Atrium. PA: Pulmonary Artery

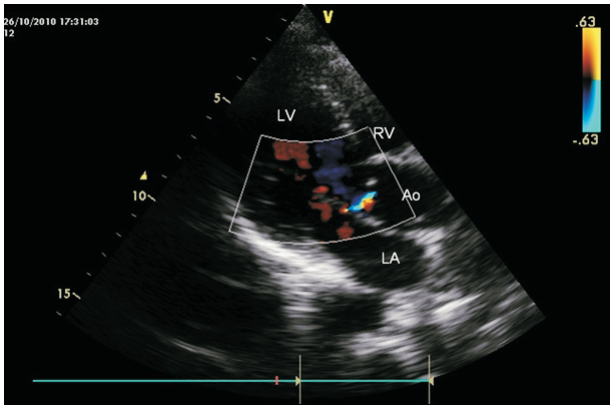

Fig. B An eccentric mild aortic regurgitation in the long axis view RV: Right Venticul, Ao: Aort, LA: Left Atrium. LV: Left Venticul

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