Asymptomatic quadricuspid aortic valve

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A 45-year-old man with isolated hypertension underwent routine echocardiography. Transthoracic examination revealed left ventricular concentric hypertrophy, mild tricuspid regurgitation and images suggestive of a bicuspid aortic valve. Transesophageal examination revealed a quadricuspid aortic valve (QAV) with normal coronary origin (four sinuses of Valsalva and four aortic cusps), x-configuration, a Greek cross appearance in diastole (Figure 1) and a rhomboidal shape in systole (Figure 2). Mild aortic regurgitation was present, but had no hemodynamic significance. Considering the absence of symptoms and valvular lesions, we recommended only endocarditis prophylaxis.

QAV is a very uncommon congenital anomaly, with an incidence of 0.008% to 0.033% (1) and equal sex distribution (2). QAV is usually diagnosed during autopsy or vascular surgery, and less frequently diagnosed with echocardiography or aortography (1). QAV may be found as an isolated lesion or in association with aortic valvular disease, usually regurgitation. In the literature, we found associations with other congenital anomalies such as ventricular septal defect (2), hypoplasia of the anterior mitral leaflet, subaortic fibromuscular

stenosis, patent ductus arteriosus (2), pulmonary artery stenosis (3), supraventricular arrhythmias (4), complete atrioventricular block and coronary artery anomalies (2,4). Hurwitz and Roberts (2) classified QAV into seven anatomical variations related to cusp dimensions. Asymmetrical QAV appears to be more associated with valvular disease (regurgitation or stenosis) as a consequence of turbulence flow through the valve and shear stress, leading to fibrotic thickening of the cusps (1).

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Figure 1) Quadricuspid aortic valve in diastole

Figure 2) Quadricuspid aortic valve in systole

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