A "4 leaf clover aortic valve" – not as lucky as it sounds

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Introduction: Qvadricuspid aortic valve (QAV) represents an extremely rare congenital heart disease with an incidence of 0.01-0.04%. It is generally an isolated anomaly, but 18-32% of patients present with another congenital heart disease.[1,2] QAV is frequently associated with progressive aortic regurgitation (AR) with the development of significant valvulopathy in the fifth to sixth decade. There are a few classifications of QAV based on leaflet size and distribution.[1-3] Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) are the main methods of QAV detection. Computed tomography (CT) may accurately show the status of QAV, dimensions of the aorta, and the location of coronary ostia.[2,3]

Case report: A totally asymptomatic 49-year-old woman was referred to our echo lab after an accidental finding of a diastolic heart murmur. A cardiovascular examination did not show signs of heart failure. TTE showed normal left ventricular dimensions, wall thickness, normal 3D-derived left ventricle volumes and ejection fraction (60%). The size of the aortic root was normal, and ascending aorta was slightly dilated. The parasternal short-axis view showed an aortic valve characterized by 4 cusps of equal size and moderate aortic regurgitation with a central aortic jet, without any further anomalies. She did not agree to the proposed TEE, but the acoustic window on TTE was accurate to see closely the aortic valve. We did a CT of the aorta and aortic valve, which showed the QAV without calcification, normal measurements of the aortic annulus and an annulus circumference, as well as the normal distance of the annulus from the coronary arteries. The ascending aorta was slightly dilated.

Conclusion: A QAV is a rare congenital heart disease whose clinical manifestations depend on the functional status of the valve and the presence of any associated anomalies. The presence of severe aortic regurgitation, severe aortic stenosis, or QAV with valvular dysfunction associated with other clinically significant abnormalities are indications for surgery.[1,2] The surgical options are replacement or valve repair. Aortic valve tricuspidalization is the most common repair technique.[1-3] Our patient was totally asymptomatic with normal ventricular function and moderate aortic regurgitation, so further TTE follow-up is indicated in this case.

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LITERATURE

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