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Quadricuspid Aortic Valve

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Abstract

Quadricuspid aortic valve is a very rare congenital abnormality with a frequency of about a frequency of 0.006% in large echocardiographic database. Most common functional abnormality is dominant aortic regurgitation.

Keywords: Quadricuspid aortic valve, Aortic regurgitation

Elderly female presented with paroxysmal atrial fibrillation with fast ventricular rate. Clinically she had an early diastolic murmur along the left sternal edge suggestive of aortic regurgitation. Echocardiogram showed quadricuspid aortic valve with moderate aortic regurgitation (**Figure 1** and **2**). ECG showed left ventricular hypertrophy in addition to atrial fibrillation with fast ventricular rate.

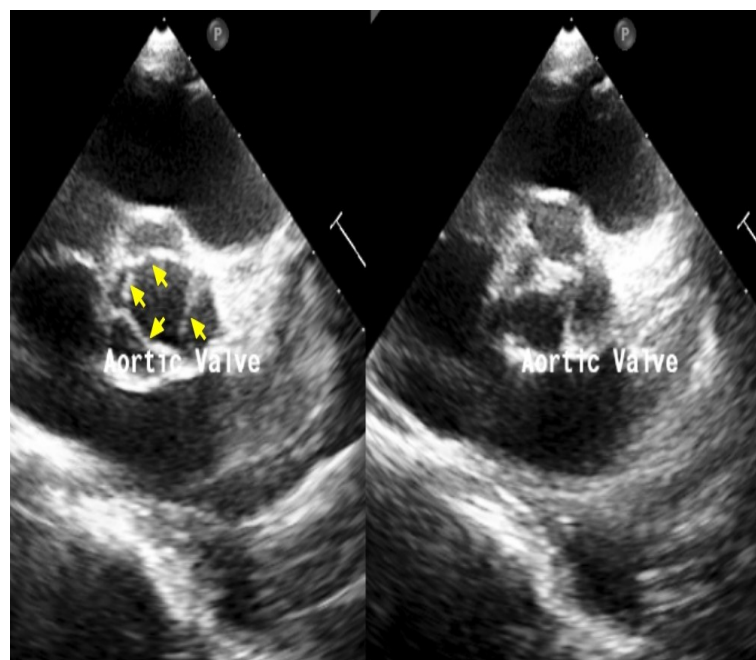


Figure 1: Quadricuspid aortic valve on echocardiogram in parasternal short axis view. Left panel shows the valve in open position with 4 cusps (yellow arrows). Right panel shows the valve in closed position.

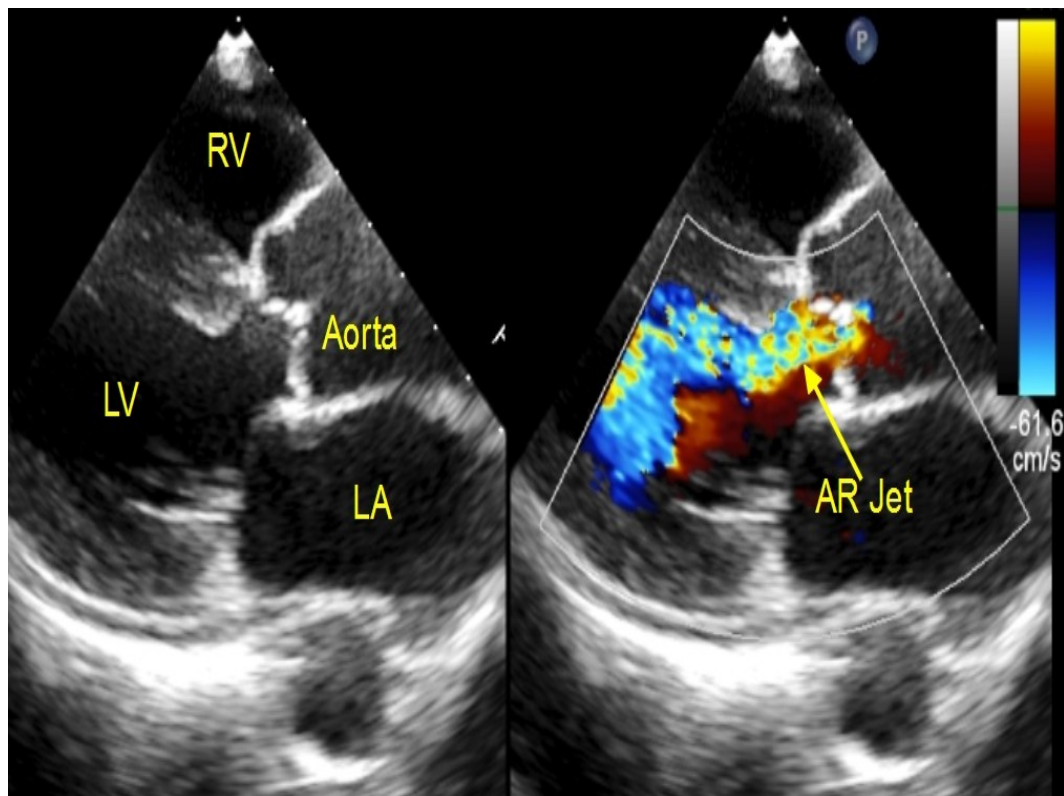


Figure 2: Colour Doppler Echocardiogram in parasternal long axis view showing moderate aortic regurgitation (AR) jet extending from the left ventricular outflow tract into the left ventricle.

Quadricuspid aortic valve is a rare congenital abnormality. Usually it produces dominant aortic regurgitation as in this case [1]. Tsang MY et al [2] evaluated a large echocardiography database of 788,733 transthoracic echocardiograms from Mayo Clinic and documented 49 cases of quadricuspid aortic valves. Another patient thought to have trileaflet aortic valve with severe aortic regurgitation was found to have quadricuspid aortic valve at surgery. Retrospective review of the echocardiographic images showed type F quadricuspid aortic valve as described by Hurwitz LE and Roberts WC [3]. So total 50 cases from the Mayo Clinic database corresponds to a frequency of 0.006% [2].

Hurwitz and Roberts classification [3] was originally developed for quadricuspid pulmonary valve which is commoner. The types are as follows:

Type A: Four equal sized cusps

Type B: Three equal cusps and one small cusp

Type C: Two equal large cusps and two equal small cusps

Type D: One large, two intermediate and one small cusp

Type E: Three equal and one larger cusp

Type F: Two equal larger and two unequal smaller cusps

Type G: Four unequal cusps

The current case appears to be Type B with three equal cusps and one small cusp as seen in **Figure 3**.

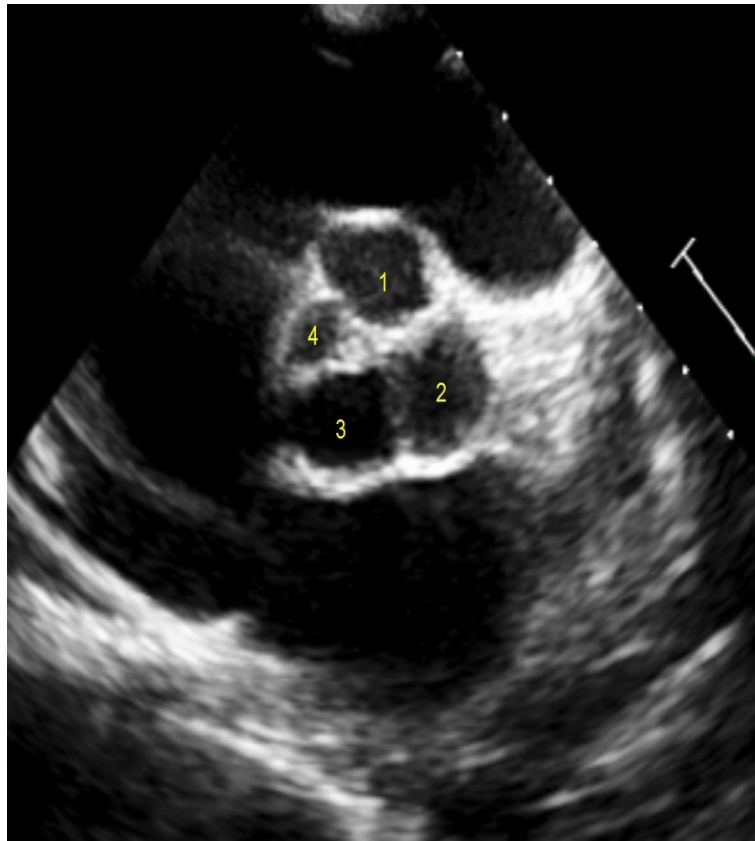


Figure 3: Quadricuspid aortic valve with three equal cusps (1-3) and one small cusp (4).

References

1. Yuan SM. Quadricuspid Aortic Valve: A Comprehensive Review. *Braz J Cardiovasc Surg.* 2016 Nov-Dec;31(6):454-460.
2. Tsang MY, Abudiab MM, Ammash NM, Naqvi TZ, Edwards WD, Nkomo VT, Pellikka PA. Quadricuspid Aortic Valve: Characteristics, Associated Structural Cardiovascular Abnormalities, and Clinical Outcomes. *Circulation.* 2016 Jan 19;133(3):312-9.
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