Quadricuspid aortic valve and double chambered right ventricle: a rare combination

▶ An additional supplementary video is published online only. To view this file please visit the journal online (http://dx.doi.org/10.1136/heartasia-2012-010203).

A 28-years-old woman presented with dyspnoea for the last 18 months. On examination, her heart rate was 90 beats/min, blood pressure 100/70 mm Hg. Jugular venous pressure was raised with prominent 'a' waves. Precordial examination revealed grade 5/6 ejection systolic murmur in left 2nd and 3rd intercostal space. The 12-lead ECG revealed right ventricular hypertrophy with right axis deviation. Two-dimensional echocardiographic and Doppler examination revealed quadricuspid aortic valve in systole and diastole and double-chambered right ventricle (DCRV) with gradient across an anomalous shelf of ~150 mm Hg (figures 1 and figures 2; S Video).

Quadricuspid aortic valve is a very rare congenital heart disorder with an incidence of 0.003–0.043% of all congenital heart disorders.[1] It usually appears as an isolated congenital anomaly but may also be associated with other malformations but never been reported with DCRV. The most commonly associated congenital malformation with quadricuspid aortic valve is coronary artery anomalies seen in ~10% of cases whereas DCRV with an incidence of ~1% of all congenital heart disorders is associated with perimembranous ventricular septal defect in ~75% of cases. The quadricuspid valve has been classified into seven types based on leaflet size and is important clinically because it may lead to aortic regurgitation in adulthood which may require surgical intervention.[2] In DCRV, surgical intervention should be considered in symptomatic patients or having peak gradients >50 mm Hg. Percutaneous balloon dilatation or alcohol ablation of the conal branch of the right coronary artery has been reported and should be reserved in patients who are not otherwise surgical candidates.

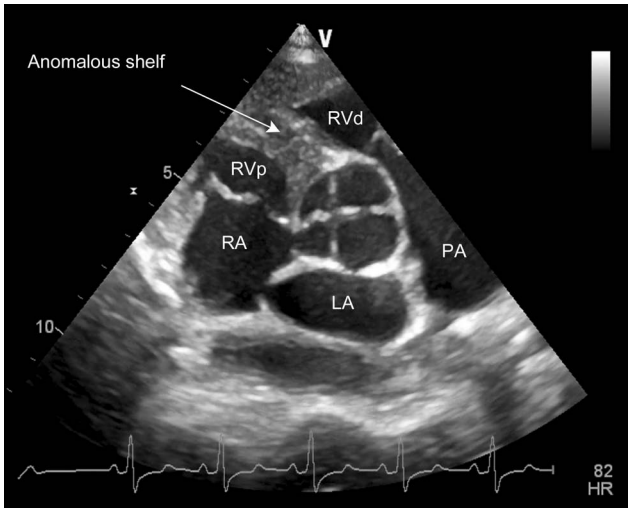

Figure 1 Transthoracic 2-D echocardiogram in parasternal short-axis view in diastole revealing quadricuspid aortic valve and anomalous shelf in right ventricle.

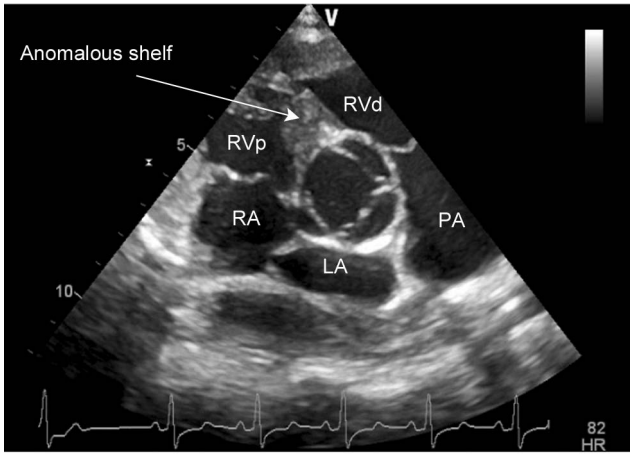

Figure 2 Transthoracic 2-D echocardiogram parasternal short-axis view in systole revealing quadricuspid aortic valve and anomalous shelf in right ventricle.

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