

A rare cause of aortic regurgitation

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A 29-year-old female patient was referred to our cardiac outpatient clinic for evaluation of a murmur. She had grade 2 symptoms of dyspnoea. On clinical examination, she had a displaced heaving apex, collapsing pulse and Duroziez's sign, all in keeping with chronic severe aortic regurgitation (AR). On transthoracic echocardiography, the parasternal long axis view showed a dilated left ventricle with mild concentric left ventricular hypertrophy (LVH). Evaluation of the aortic valve

revealed severe AR with a quadricuspid aortic valve. Further imaging of the aortic valve with transoesophageal echocardiography demonstrated bowing of the leaflets (Panel A), which usually suggests either a bicuspid aortic valve or rheumatic aortic valve disease. Morphological evaluation of the valve demonstrated 4 cusps of equal size with a central area of none coaptation as cause for severe AR (Panels B - D).

Quadricuspid aortic valve (QAV) is a rare cause of congenital aortic regurgitation with an incidence of 0.008 - 0.043% on



PANEL B: TEE short axis of the aortic valve demonstrating 4 cusps with a central defect during diastole.

CLICK TO PLAY VIDEO CLIP OF PANEL A

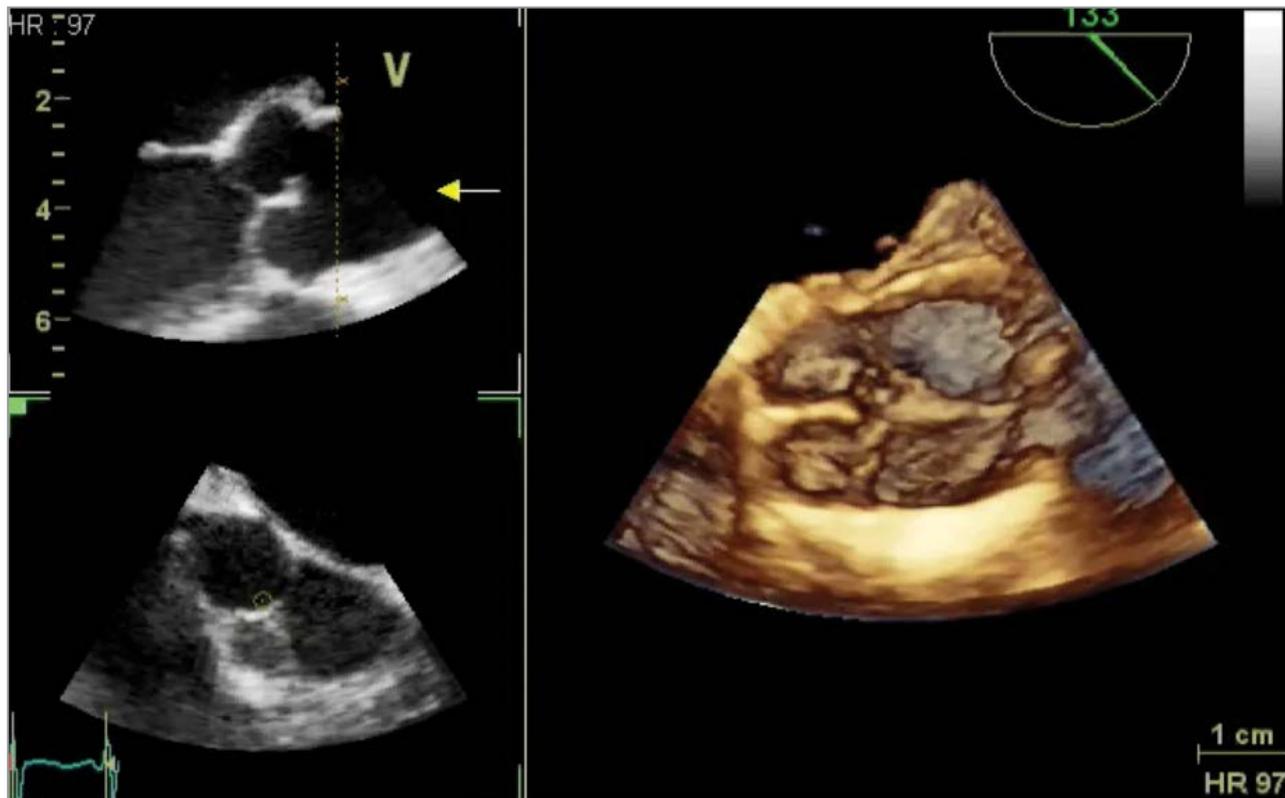


PANEL A: Transoesophageal echocardiogram (TEE) of aortic valve in long axis demonstrating bowing of the leaflets in systole.

CLICK TO PLAY VIDEO CLIP OF PANELS B AND C



PANEL C: TEE short axis of the aortic valve.



PANEL D: 3 dimensional short axis view of aortic valve.

echocardiography and 1% at surgery.⁽¹⁾ Several theories have been advanced to explain the etiology including aberrant fusion of aortopulmonary trunk of the foetal heart, abnormal proliferation of cusps or formation of an extra cusp. There are 7 anatomic subtypes depending on size of cusps, as classified by Hurwitz and Roberts.⁽¹⁻³⁾ Functionally, 75% of patients present with AR, 9% have both AR and AS, and 16% have a normal functioning valve.^(1,4)

QAV is also associated with other congenital anomalies such as abnormal origin of coronary arteries, ventricular septal defects, patent ductus arteriosus, pulmonary stenosis and hypertrophic cardiomyopathy.^(1,5) Our patient had no associated congenital anomalies.

Conflict of interest: none declared.

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