An incidentally found unusual quadricuspid pulmonary valve with transthoracic echocardiography

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A 76-year-old female was admitted to the hospital with progressive dyspnoea. There was marked cardiomegaly and diffuse pulmonary oedema on chest X-ray and pitting oedema. On admission, transthoracic echocardiography revealed large amount of pericardial effusion with haemodynamic compromise (early diastolic right ventricular collapse, early systolic right atrial inversion, respiratory variation of mitral E wave, and plethora of inferior vena cava) and moderate pulmonary regurgitation (Figure 1B) with dilated main pulmonary artery (Figure 1A). Additionally, echocardiography revealed an unusual quadricuspid pulmonary valve in the parasternal long-axis view (Figure 1C–F). The pulmonary valve has unusual four even-sized cusps, with poor coaptation and pulmonary regurgitation. Pericardial fluid cytology revealed malignant lymphoma, diffuse large B cell type.

Quadricuspid pulmonary valve is a rare congenital anomaly and occasionally combined with pulmonary regurgitation. Diagnosis of a quadricuspid pulmonary valve by transthoracic two-dimensional echocardiography is very difficult due to the anatomical disposition of the valve. Because of its rare association with significant clinical complications or symptoms, quadricuspid pulmonary valve has been diagnosed almost exclusively after death.