Quadricuspid aortic valve: an unusual cause of aortic regurgitation in rheumatoid arthritis

A 44-year-old woman with long-standing rheumatoid arthritis presented with exertional breathlessness since the last 3 months; cardiovascular examination revealed a decrescendo diastolic murmur along the left upper sternal border. A transthoracic and transesophageal echocardiogram revealed normal sized left ventricle, normal ventricular systolic function and a quadricuspid aortic valve (QAV) with mild aortic regurgitation (figure 1A,B; supplementary video 1). A 3D echocardiographic reconstruction was performed which clearly delineated the anatomy of the quadricupid aortic valve with four equal sized cusps forming the characteristic 'X' configuration during diastole (figure 1C) and opening fully during systole (figure 1D; supplementary video 2). Mild central leaflet tip mal-coaptation with AR was confirmed.

Although previously reported at autopsy or during cardiac surgery, the advent of sophisticated echocardiography techniques has resulted in cases of QAV being reported more frequently.¹ Seven anatomic variations of QAV are described, with the type having four equal cusps (type A) being the most frequent, as observed in our case.² While aortic regurgitation is common due to cuspal mal-coaptation, aortic stenosis is rare.

Associations of QAV include hypertrophic cardiomyopathy, atrial septal defect, patent ductus arteriosus, ventricular septal

defects, bicuspid pulmonary valve and congenital coronary anomalies. Rheumatoid arthritis often leads to aortic regurgitation (AR) due to aortitis with dilated aortic annulus, aortic valve prolapse or inflammatory involvement of the valve itself.³ To the best of our knowledge, presence of QAV leading to AR has not been reported before in rheumatoid arthritis.

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Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; internally peer reviewed.

► Additional material is published online only. To view please visit the journal online (http://dx.doi.org/10.1136/heartasia-2013-010349).

To cite Sharma J, Kapoor A, Kumar S. *Heart Asia* Published Online First: [*please include* Day Month Year] doi:10.1136/heartasia-2013-010349

Heart Asia 2013;0:98. doi:10.1136/heartasia-2013-010349

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Figure 1 Transthoracic (A) and transesophageal echocardiogram (B) revealed a quadricuspid aortic valve (QAV) with mild aortic regurgitation. A 3D echocardiographic reconstruction demonstrated a type A variety of QAV with four equal sized cusps forming the characteristic 'X' configuration during diastole (C) and opening fully during systole (D).