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# A rare case of quadricuspid aortic valve with moderate aortic regurgitation with left ventricular non compaction presenting with ventricular tachycardia

Arnab Ghosh Chaudhury*, B. Prabhavathi, C.N. Manjunath

*Sri Jayadeva Institute of Cardiovascular Sciences & Research, Bangalore, India*

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ABSTRACT

Quadricuspid aortic valve is a rare entity. Left ventricular noncompaction is an uncommon cause of cardiomyopathy. Combination of these two conditions is very rare. We report a case of quadricuspid aortic valve and moderate aortic regurgitation with left ventricular noncompaction who had presented with ventricular tachycardia.



## Introduction

Quadricuspid aortic valve (QAV) is a rare congenital cardiac defect with an estimated frequency of <0.05%.[1] the characteristics, natural history, and long-term outcomes of QAV are poorly defined because of its rarity. Left ventricular noncompaction (LVNC) is an anatomic variant of left ventricular structure, a rare entity with prevalence of 0.014% in adults.[2] We report a case of 35-year-old male with QAV and moderate aortic regurgitation (AR) with LVNC who had presented with ventricular tachycardia (VT).

## Case report

A 35-year-old male patient presented with hemodynamically unstable monomorphic VT. He was treated with DC cardioversion and was stabilized. Patient had history of regular exertional palpitation for 1 year before this episode without any significant family history. Physical examination revealed early diastolic murmur of short duration in neoaortic area, without any evidence of aortic run off. Routine blood investigations including serum electrolytes were normal. Electrocardiogram showed sinus rhythm, left atrial enlargement and left ventricular hypertrophy. Chest X-ray showed cardiomegaly. Echocardiography revealed dilated left atrium, left ventricle with global left ventricular hypokinesia (ejection fraction [EF] 32%) and moderate aortic regurgitation (AR) (Fig. 1A). Severe AR was ruled out as there was no holodiastolic reversal of flow in descending aorta. Vena contracta was 5 mm. In parasternal short axis view QAV was identified (Figs. 1C, 2A) (one large, two intermediate, one small sized cusp-Hurwitz and Roberts type D).[3] On further evaluation, trabeculations and intertrabecular recesses were identified predominantly involving apicolateral and inferior left ventricular (LV) walls (Fig. 1B). Cardiac MRI showed LV trabeculations in apicolateral and inferior segments with deep intertrabecular recesses (Fig. 2B) with noncompacted: compacted LV wall ratio 2:1. It also revealed thinned out and dyskinetic apical anterior segment and true apex (Fig. 2C) suggestive of left anterior descending (LAD) artery territory infarct. Coronary angiography was done. There was no flow limiting lesion in epicardial coronaries.

## Discussion

Quadricuspid aortic valve is a rare congenital cardiac defect (frequency 0.006%).[4] Morphologically it is classified by Hurwitz and Roberts in 6 types A to F according to relative sizes of the cusps. Our case had type D aortic valve (one large, two intermediate and one Small sized cusp). The 3 commonest QAV subtypes are types A, B, and C. Patients present predominantly with AR in 26% of cases while aortic stenosis (AS) is seen rarely in about 8% of cases. Progressive cusp fibrosis with subsequent failure of cusp coaptation over time has been suggested as the key mechanism in regurgitation. There is no association between morphological type of aortic valve cusp and severity of AR. Associated aortic root dilatation is com-

* Corresponding author at: 122/50 Kumara Krupa JP Nagar 3rd Main 3rd Phase, Bangalore, Karnataka, 560078, India.
*E-mail addresses:* drarnabgc@rediffmail.com, arnab@jayadevacardiology.com (A.G. Chaudhury), prabhavathi_in@yahoo.com, prabhavathi@jayadevacardiology.com (B. Prabhavathi), director@jayadevacardiology.com (C.N. Manjunath).

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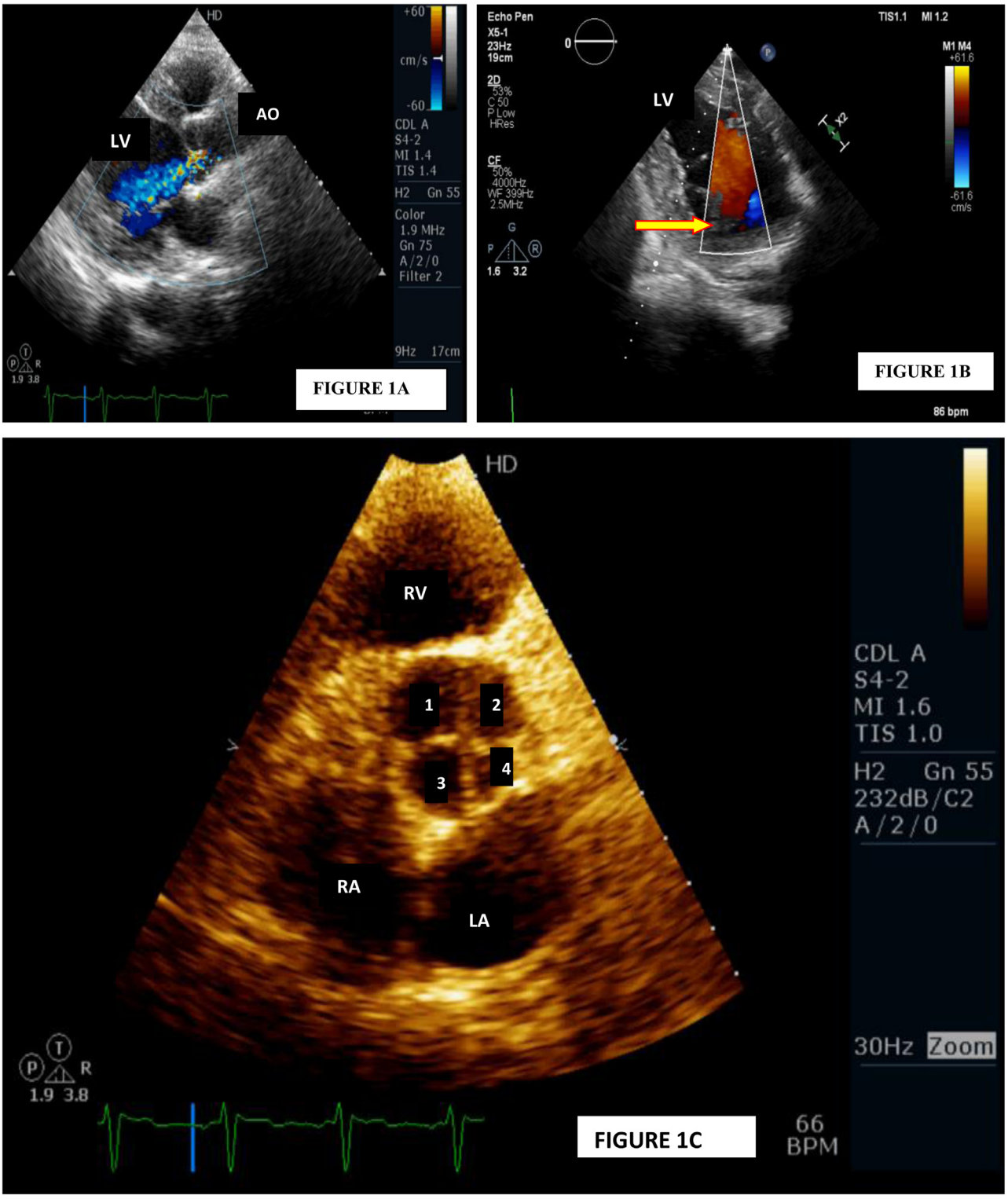


**Fig. 1.** (A) parasternal long axis view showing moderate aortic regurgitation. LV: left ventricle; AO: aorta. (B) modified parasternal short axis view showing prominent trabecular recesses filling with blood (yellow arrow). LV: left ventricle. (C) parasternal short axis view at aortic valve level showing quadricuspid aortic valve. one large(1), two intermediate(2,3), one small(4) sized cusps- Hurwitz and Roberts type D. LA: left atrium; RA: right atrium; RV: right ventricle.

mon (29%) but infective endocarditis or aortic dissection is rare. Aortic regurgitation is slowly progressive, rarely requires surgery (aortic valve repair or replacement). Quadricuspid aortic valve is generally an isolated anomaly, rarely associated with other congenital lesions. Long-term survival is excellent both without and with aortic valve surgery.[4] LVNC is defined by 3 markers: prominent left ventricular trabeculae, deep intertrabecular recesses, and the thin compacted layer (noncompacted to compacted ratio 2:1 or more). Ventricular tachyarrhythmias have been reported in up to 47% of symptomatic patients. Sudden cardiac death (SCD) has been reported in 13% to 18% of (mostly adult) patients with LVNC. LVNC can be isolated or associated with congenital heart diseases.