

*Images in Cardiovascular Medicine*

# Nonischemic Dilated Cardiomyopathy With Quadricuspid Aortic Valve

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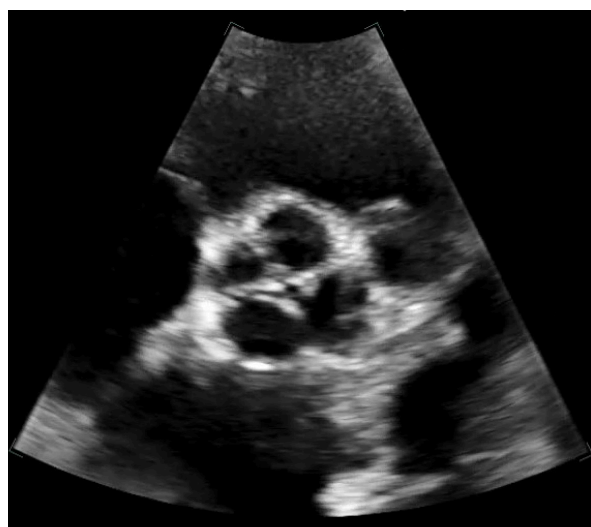
## Case Description

A 54-year-old man with a history of HIV, substance use disorder, and hyperlipidemia presented at the emergency department with dyspnea worsening on exertion and lower-extremity edema. Electrocardiogram showed a new-onset bifascicular block, whereas the echocardiogram revealed a 24% left ventricular ejection fraction and a 4-leaf clover appearance of the aortic valve, with moderate to severe regurgitation. A quadricuspid aortic valve was discovered (Fig. 1).

## Comment

The quadricuspid aortic valve, which resembles a 4-leaf clover on a transthoracic echocardiogram, is a rare congenital cardiac condition seen in 0.0059% to 0.0065% of patients.<sup>1</sup> The majority of cases often appear in the fifth decade of life, with varying degrees of aortic regurgitation. Reverse blood flow to the left ventricle is facilitated by the uneven blood shear force on 4 rather than 3 valves. Backflow that is left untreated for a long period remodels the ventricle to accept the extra jet, resulting in dilated cardiomyopathy. Surgical repair rather than valve replacement is performed on an individual patient basis, typically saved for situations with significant cardiac insufficiency.

Despite receiving a full explanation of the treatment plan and tricuspidation technique, this patient declined further interventions. Stabilized heart failure with reduced ejection fraction was achieved medically; the patient also underwent implantable cardioverter-defibrillator placement. Notably, no ventricular perfusion abnormalities were detected during a nuclear stress test.



**Fig. 1** Transthoracic echocardiogram, short axis view, shows the 4-leaf clover appearance of the aortic valve, compatible with a quadricuspid valve anomaly.

Supplemental motion image is available for [Figure 1](#).

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## References

1. Tsang MYC, Abudiab MM, Ammash NM, et al. Quadricuspid aortic valve: characteristics, associated structural cardiovascular abnormalities, and clinical outcomes. *Circulation*. 2016;133(3):312-319. doi:10.1161/CIRCULATIONAHA.115.017743