Quadricuspid Aortic Valve in a Young Adult: Lessons from Serial Echocardiograms

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Abstract:

Background: Quadricuspid aortic valve (QAV) is an extremely rare congenital cardiac anomaly, with a reported incidence of less than 0.05%. It is frequently misdiagnosed as a trileaflet aortic valve on transthoracic echocardiography. Recognition is clinically important due to its association with aortic regurgitation (AR), endocarditis, and surgical implications.

Case Report: A 21-year-old male presented for a routine health check-up. Echocardiography revealed a QAV with mild AR, preserved biventricular function, and no aortic stenosis. He had a history of prolonged febrile illness at age 13, later diagnosed as Epstein Barr virus associated infectious mononucleosis. At that time, echocardiography demonstrated a trileaflet aortic valve with trivial AR and no vegetation. Serial echocardiograms between 2018 and 2022 repeatedly reported a trileaflet morphology with trivial to mild AR. The correct diagnosis of QAV was established only in 2024, when parasternal short-axis imaging clearly demonstrated four cusps. The patient was advised to have endocarditis prophylaxis and regular echocardiographic surveillance.

Conclusion: This case underscores the diagnostic challenges of QAV and highlights the importance of meticulous echocardiographic evaluation, as accurate recognition impacts long-term surveillance, counselling, and surgical planning.