Images in Cardiovascular Medicine

Quadricuspid Aortic Valve

Revealed by Real-Time, 3-Dimensional Transesophageal Echocardiography

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A 43-year-old woman presented with a long history of increasing dyspnea upon exertion. Examination revealed a diastolic aortic murmur and a blood pressure of 150/75 mmHg. The patient had undergone several transthoracic echocardiographic (TTE) examinations in the last 5 years; all had revealed only moderate aortic regurgitation. Two-dimensional transesophageal echocardiography (TEE) (Figs. 1 and 2), and real-time 3-dimensional TEE with use of a Philips iE33 system and X7-2t TEE matrix transducer (Philips Electronics N.V.; Best, The Netherlands), showed a quadricuspid aortic valve (QAV) associated with moderate-to-severe aortic regurgitation (Figs. 3 and 4). Left ventricular size and function were preserved. Computed tomography revealed a hypoplastic left brachiocephalic vein. The patient declined further evaluation for surgery.

Comment

Quadricuspid aortic valve is an uncommon congenital anomaly; the reported prevalence on autopsy is about 0.01%.[1] Benjamin Guy Babington was apparently the first to note a QAV (April 1847),[2] and that autopsy case was reported in 1848.[3] Al-

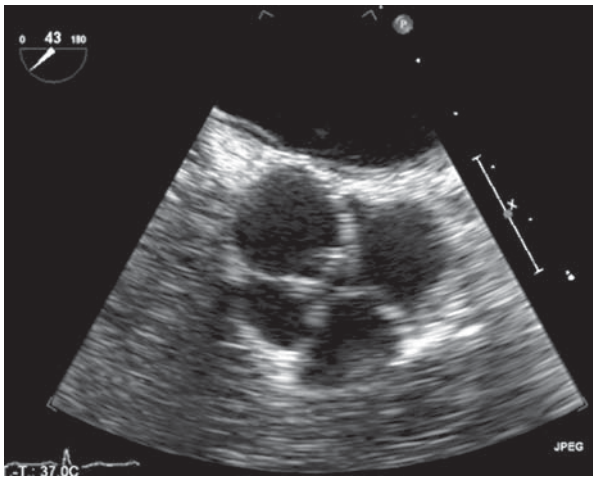

Fig. 1 *Transesophageal echocardiogram (2-dimensional short-axis view) shows an aortic valve with 4 cusps.*

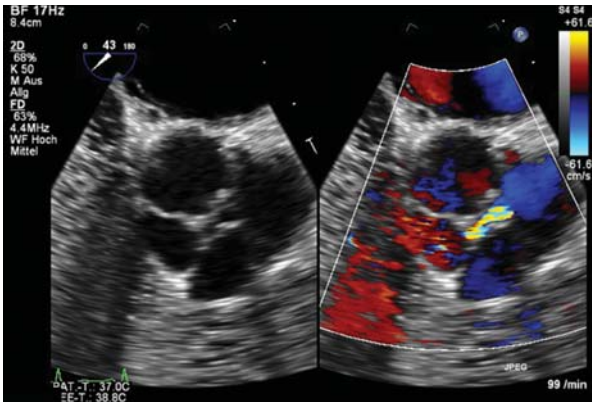

Fig. 2 *Transesophageal echocardiogram (2-dimensional short-axis view) shows the quadricuspid aortic valve. At right, color flow indicates aortic regurgitation.*

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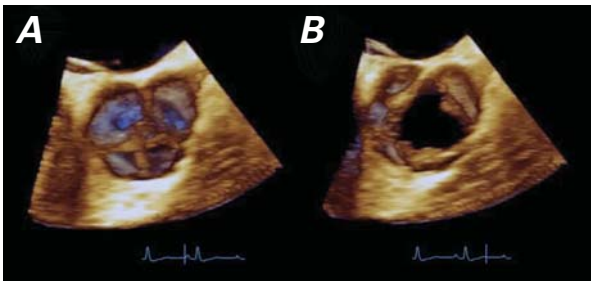

Fig. 3 *Transesophageal echocardiograms (3-dimensional short-axis views) show the* ***A****) closed and* ***B****) open quadricuspid aortic valve.*

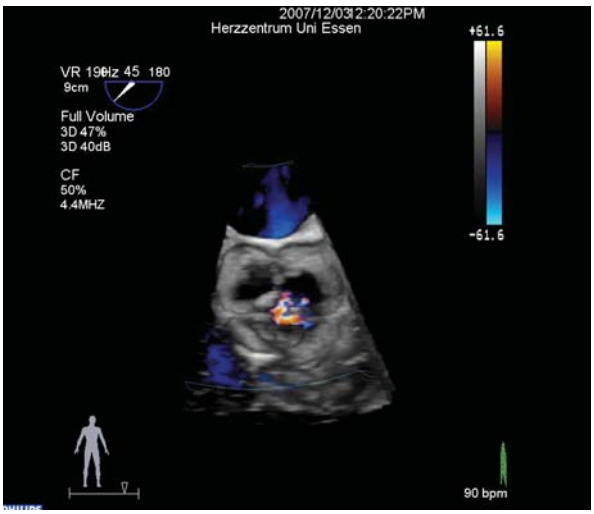

Fig. 4 *Transesophageal echocardiogram (3-dimensional view with color flow) shows the quadricuspid aortic valve.*

though several anatomic variations of QAV have been described, there are typically 3 cusps of equivalent size and a small 4th cusp between the right coronary cusp and the noncoronary cusp. In accordance with cusp sizes, QAVs are classified into 7 types, named A to G.[4] Our patient's valve had 3 equal-sized cusps and a smaller accessory cusp (type B). In a review of QAV cases,[1] 75% of patients also presented with aortic regurgitation; other malformations included an abnormally located coronary ostium, pulmonary valve stenosis, nonobstructive cardiomyopathy, subaortic stenosis, and ventricular septal defect.[1] Therefore, thorough examination of the patient and close follow-up of the aortic regurgitation are necessary. When the cause of significant aortic regurgitation is not apparent, TEE—particularly with 3-dimensional imaging—can be helpful.

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