

IMAGES IN CLINICAL MEDICINE

Uni-, Bi-, Tri-, and Quadricuspid Aortic Valves

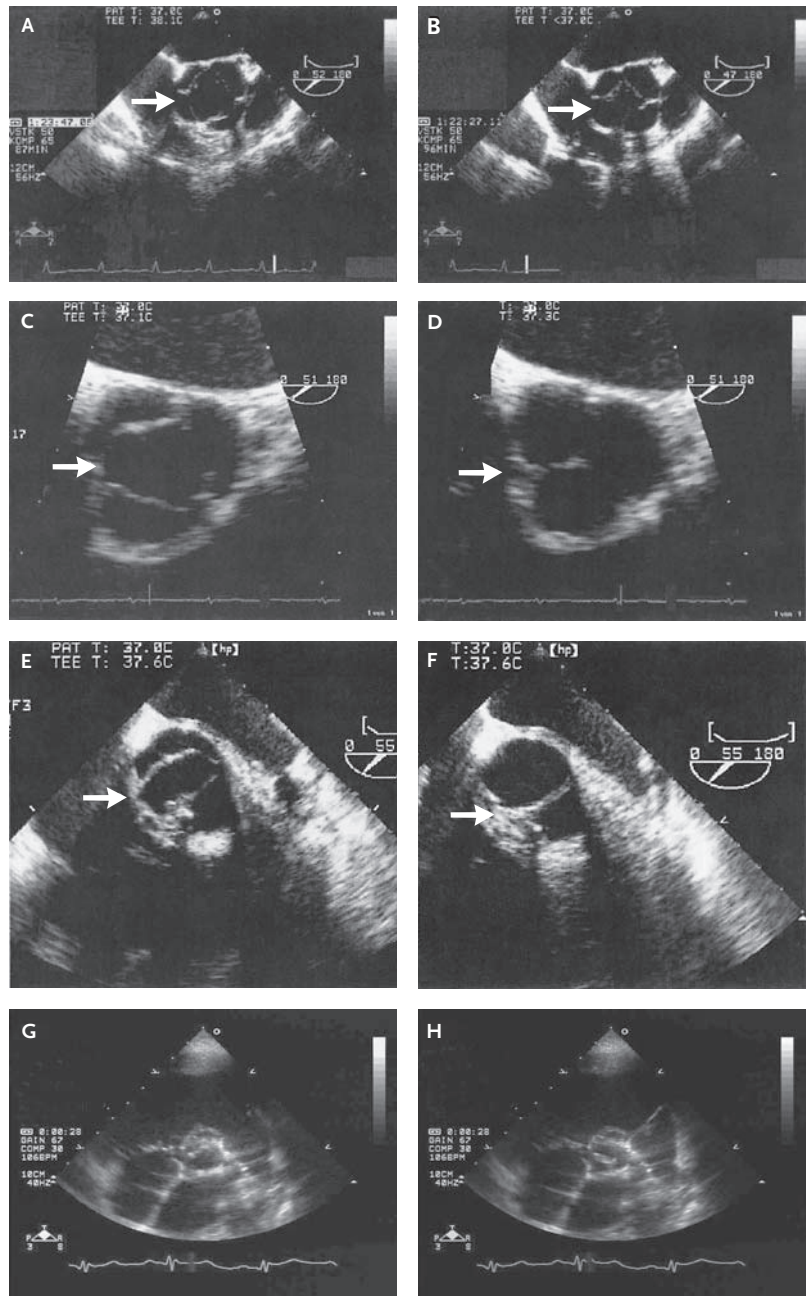
A 47-YEAR-OLD WOMAN PRESENTED with a 6-month history of left-sided chest pain and minimal dyspnea on exertion. On physical examination, her blood pressure was 120/60 mm Hg and a diastolic murmur of grade 2/6 to 3/6 was heard at the left sternal border. The results of electrocardiography were normal. Echocardiography showed a quadricuspid aortic valve with four slightly calcified and thickened cusps of equal size (arrows, Panel A in systole and Panel B in diastole; for comparison, a normal tricuspid aortic valve is shown, Panel C in systole and Panel D in diastole). This malformation was associated with moderate-to-severe aortic regurgitation. Left-heart catheterization showed normal coronary arteries with a displaced left main stem but no other cardiac abnormalities.

A bicuspid aortic valve (arrows, Panel E in systole and Panel F in diastole) is one of the most common congenital malformations of the heart and is associated with an increased risk of endocarditis and aortic stenosis. A unicuspid aortic valve (Panel G in systole and Panel H in diastole) occurs rarely and is typically diagnosed in early childhood in association with aortic stenosis. A congenital quadricuspid valve is a rare cause of pure aortic regurgitation and can be associated with displacement of the coronary ostium, indicating a developmental defect. This patient's symptoms were caused by aortic insufficiency, and she was treated with medical therapy. Clinical and echocardiographic follow-up at 1 year showed that the patient was in stable condition.

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Panels G and H courtesy of Dr. Amy L. Juraszek, Children's Hospital, Boston.