

Unicuspid aortic valve concomitant with interrupted aortic arch and severe aortic stenosis in an adult patient

Yetişkin hastada aortik ark kesintisi ve ileri aort darlığının eşlik ettiği uniküspit aortik kapak

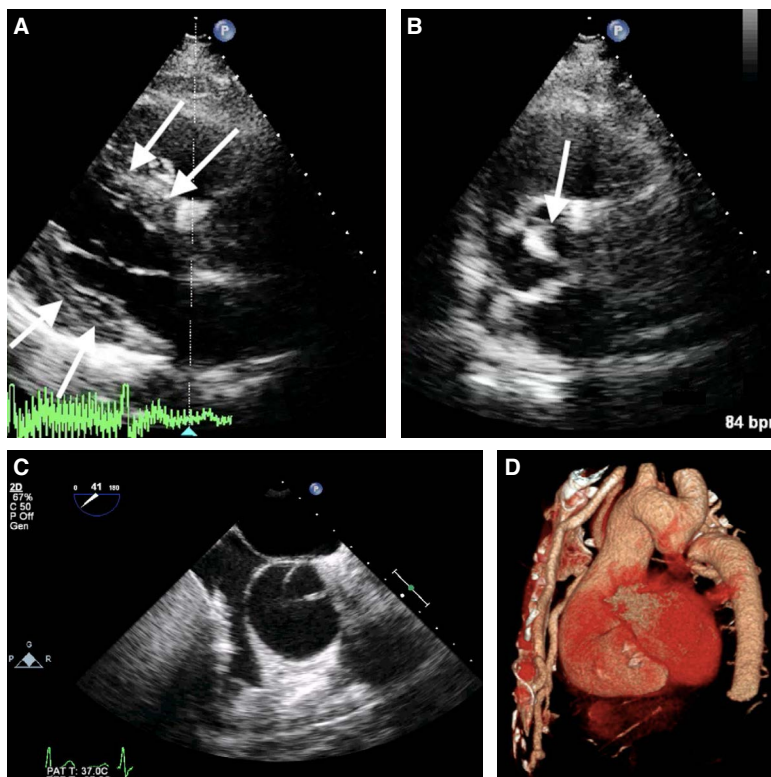
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A 41-year-old male patient presented at the cardiology clinic with progressively increasing, squeezing chest pain. His medical history was significant for hypertension diagnosed a year earlier. The physical examination results indicated that while his blood pressure was approximately 165/95 mm Hg in the upper extremities, it decreased to

110/75 mm Hg in the lower extremities. Cardiac auscultation revealed a grade 4/6 mid-systolic ejection murmur. Transthoracic echocardiography demonstrated calcific aortic valve morphology and aortic stenosis with a peak gradient and mean gradient of 68 mm Hg and 37 mm Hg, respectively (Figure A, B). During a scheduled preoperative coronary angiography, a guidewire could not be advanced to the aortic

arch via the femoral approach and the descending thoracic aortography showed complete occlusion of the descending thoracic aorta compatible with an interrupted aortic arch (Video 1*). Due to severe tortuosity of the right and left subclavian arteries, cannulation of the coronary arteries could not be completed and only ascending aortography was performed (Video 2*). Transesophageal echocardiography demonstrated severe stenosis of a unicuspid unicommisural aortic valve associated with interruption between the distal aortic arch and the proximal descending thoracic aorta (Figure C, Video 3*). In order to clarify the aortic anatomy, contrast-enhanced computed tomographic angiography was performed, which confirmed interruption of the descending thoracic aorta just distal to the left subclavian artery (Figure D). Although many abnormalities associated with unicuspid unicommisural aortic valve have been described, to the best of our knowledge, this is the first report of an adult patient with unicuspid unicommisural aortic valve, interrupted aortic arch, and severe aortic stenosis.



(A) The left ventricular hypertrophy as observed in the parasternal long axis of transthoracic echocardiography, and (B) the calcific aortic valve as seen in the parasternal short axis view of transthoracic echocardiography. (C) The unicuspid unicommisural aortic valve as seen in the mid-esophageal short axis view of transesophageal echocardiography. (D) The reconstructive image of the thorax designating the interrupted aortic arch, as seen on contrast-enhanced computed tomography.

*Supplementary video files associated with this presentation can be found in the online version of the journal.