

Quadricuspid aortic valve associated with persistent left superior vena cava and right ventricular noncompaction cardiomyopathy

Persistan sol süperiyör vena kava ve sağ ventriküler süngerimsi miyokart ile ilişkili dört yaprakçıklı aortik kapak

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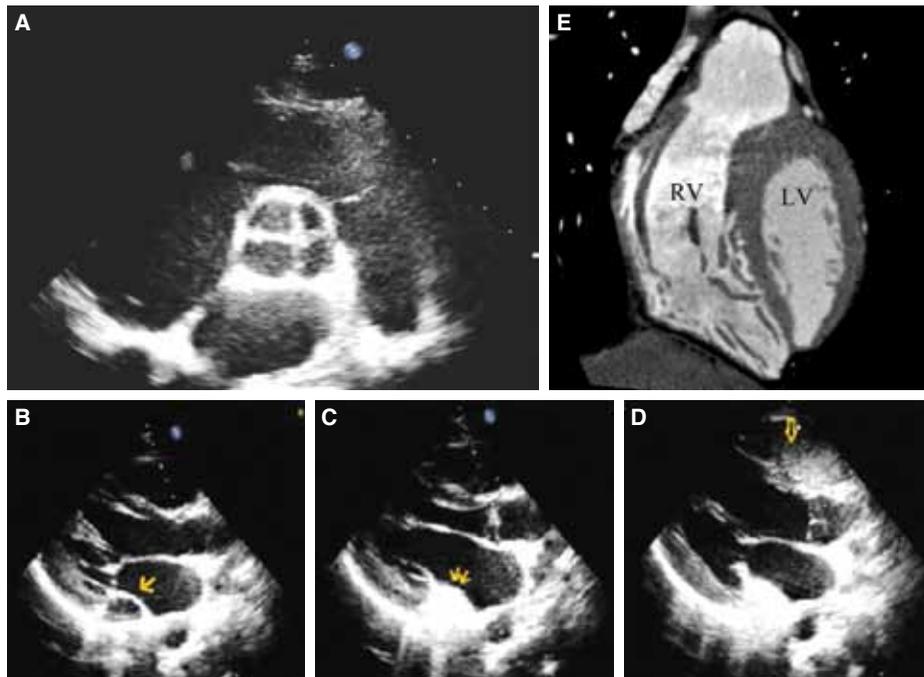
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A 27-year-old male was admitted to our department for evaluation of increasing dyspnea on exertion. He denied chest pain. On physical examination, his blood pressure was 125/85 mmHg, and an evident diastolic murmur of grade 2/4 was heard at the third left intercostal space. Results of electrocardiography and chest radiography were normal. Transthoracic echocardiography revealed a quadricuspid aortic

valve, a moderate aortic regurgitation and a dilated coronary sinus (Fig. A, Video 1*). There was no atrial or ventricular dilation, and systolic function was normal.

Owing to the dilated coronary sinus seen in the parasternal long axis, we suspected persistent left superior vena cava (Fig. B-D, Video 2*). Based on this suspicion, we injected agitated saline into the left arm. After injection, the coronary sinus opacified before the right atrium and right ventricle. The diagnosis of a persistent left superior vena cava was likely. Cardiac computed tomography (CT) was performed, after which a diagnosis of a congenital quadricuspid aortic valve associated with persistent left superior vena cava was verified. Incidentally, a right ventricular noncompaction cardiomyopathy was noted (Fig. E). No further testing was done, and the patient has done well on regular follow-up.



Figures– (A) Two-dimensional echocardiography in the parasternal short-axis view shows quadricuspid aortic valve. (B) Two-dimensional echocardiography in the parasternal long-axis view shows a dilated coronary sinus (arrow). (C) After injection of i.v. saline contrast from the left arm, opacification of the coronary sinus in the early phase (double arrow) is seen. (D) After injection of i.v. saline contrast from the left arm, opacification of right chambers in the late phase is seen (thick arrow). (E) Multi-slice computed tomography shows prominent trabeculations and deep intertrabecular recesses in the right ventricle (LV: Left ventricle; RV: Right ventricle). *Supplementary video files associated with this presentation can be found in the online version of the journal.