

## A co-anomaly: hourglass-like aorta and giant coronary arteries

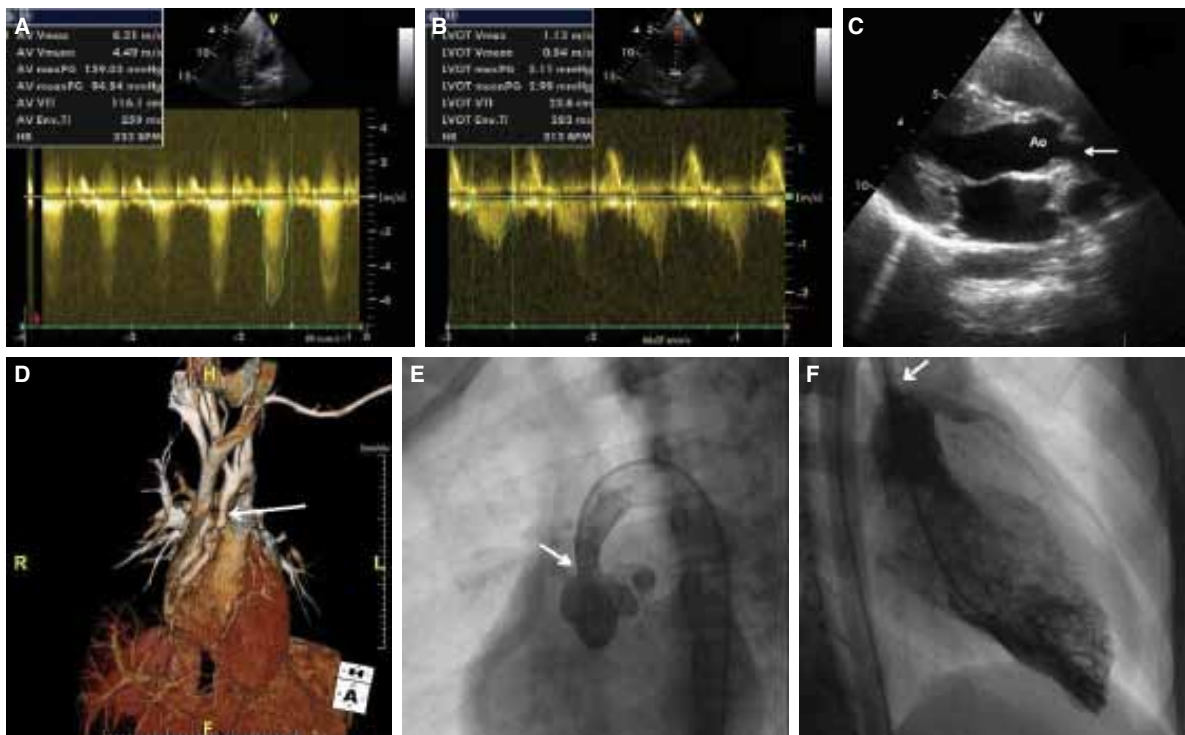
### Kum saati aort ve dev koroner arter birlikteliği

Mehmet Ata Akıl  
Mehmet Zihni Bilik  
Halit Acet,  
Faruk Ertaş  
Abdülkadir Yıldız

Department of Cardiology,  
Dicle University Faculty of  
Medicine, Diyarbakır

Supravalvular aortic stenosis (SVAS) is a rare form of congenital heart disease that occurs as a localized or diffuse narrowing of the ascending aorta beyond the sinotubular junction causing left ventricular outflow tract (LVOT) obstruction. An 18-year-old male was referred to our hospital by a cardiologist for an incidental finding of heart murmur, with the diagnosis of hypertrophic obstructive cardiomyopathy (HoCM). He had experienced atypical chest pain for a few months. His blood pressure, measured on the right and left arms, was 100/60 and 90/60 mmHg, respectively. A grade 3/6 mid-systolic murmur was heard at the second intercostal space on the right side and at the third intercostal space on the left side of the chest. He had a normal facial appearance and mental status. No other abnormalities

were found on the physical examination. The electrocardiogram showed left bundle branch block. Transthoracic echocardiography revealed a hypertrophic left ventricle with no outflow tract obstruction and SVAS with a maximum pressure gradient of 159 mmHg (Figs. A, B, C; Videos 1, 2\*). Computed tomographic angiography confirmed the diagnosis of SVAS by showing a narrowing of the aorta at the superior margin of the sinus of Valsalva with significant accelerated flow (Fig. D). Aortic root angiogram showed a narrowed segment of the ascending aorta just above the sinotubular junction (Figs. E, F; Video 3\*). A maximum pressure gradient of 120 mmHg was detected within, below and above the narrowed segment of the aorta. Selective angiography revealed aneurysmatic dilatation of both coronary arteries (Video 4\*). The SVAS was eventually corrected surgically with no complications. Misdiagnosis of HoCM can lead to improper anxiety, unnecessary medication, and further evaluation. SVAS should be kept in mind in patients with left ventricular hypertrophy.



**Figures**— Transthoracic echocardiography revealing pressure gradients from (A) supraaortic stenosis, (B) left ventricular outflow and (C) left ventricular hypertrophy and supraaortic stenosis. (D) Supravalvular aortic stenosis and giant coronary arteries in three-dimensional computed tomography. (E) Hourglass-like supravalvular aortic stenosis and giant coronary arteries. (F) Left ventriculography showing supraaortic narrowing with a normal cavity. \*Supplementary video files associated with this presentation can be found in the online version of the journal.