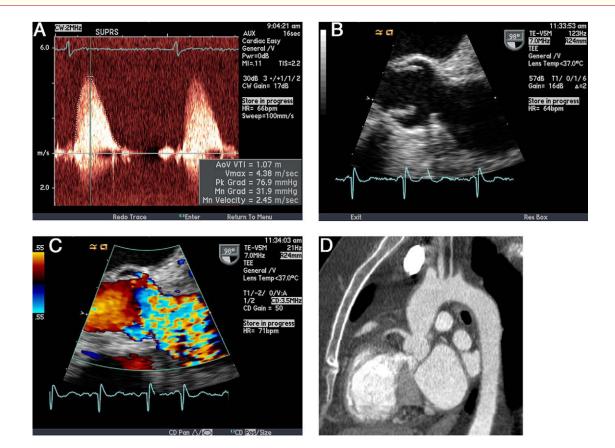
An Hourglass-Type Supravalvular Aortic Stenosis

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From the Divisions of *Cardiology and †Radiology, UMDNJ-New Jersey Medical School, Newark, New Jersey. Manuscript received April 28, 2009; revised manuscript received September 8, 2009, accepted September 14, 2009. 49-year-old woman with exertional chest pain and a systolic murmur at the right upper sternal border radiating to both carotids had a hypercontractile left ventricle (LV) and a 77 mm Hg peak pressure gradient between the LV and the ascending aorta on transthoracic echocardiography (A). Transesophageal echocardiography revealed a membrane-like intimal thickening (B) with flow acceleration at the sinotubular junction (C, Online Video 1) and no anomalies of LV outflow tract or the aortic valve. Computed tomography demonstrated hourglass-shaped aortic root with 1-cm diameter at the stenosis (D, Online Video 2). Final diagnosis was supravalvular aortic stenosis. Hyperdynamic LV likely contributed to pressure gradient. The patient declined surgery.