

Images in cardiovascular medicine

Membrane-type subaortic stenosis in the adult

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In 1992 a sinus venosus type of interatrial septal defect with a partially anomalous pulmonary venous return to the superior vena cava was diagnosed in a 46-year-old woman. The cardiac congenital abnormality was surgically repaired in 1994. In March 1998, following the finding of a crescendo-decrescendo systolic murmur

(previously not evident and possibly related to the increased left ventricular output after closure of the shunt), the patient was submitted to Doppler echocardiography which revealed the presence of mild aortic stenosis (mean transaortic gradient 20 mmHg). The mean gradient progressively increased and in May 2000 it was 50

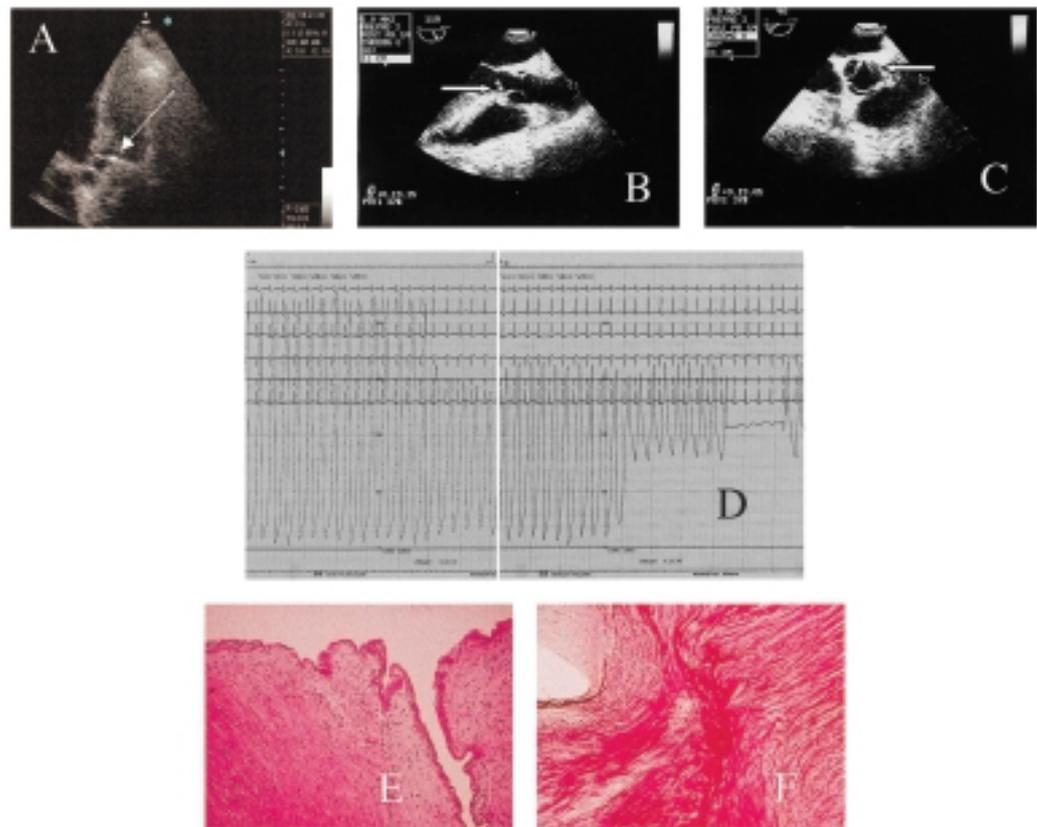


Figure 1. Transthoracic (5-chamber view) (A) and upper transesophageal (long-axis view) echo images (B) showing a spur in the left ventricular outflow tract (arrows). C upper transesophageal short-axis echo images of the normal opening (end-systole) of the aortic valve. D: left cardiac catheterization pullout pressure tracing showing an intraventricular systolic gradient. E: hematoxylin-eosin staining showing the accumulation of collagen and elastic fibers in the subendocardium. F: van Gieson staining showing the collagen and elastic fibers in the diaphragm intensely stained in red.

mmHg (triple measurement). The patient started to complain of effort dyspnea and on one occasion even fainted. In January 2001, catheterization showed normal coronary arteries. A pressure recording during the catheter pullout showed a mean systolic gradient of about 60 mmHg, consistent with another Doppler echo measurement. The images obtained at ventriculography were consistent with the presence of a subvalvular diaphragm in the left ventricular outflow tract and a hypertrophic left ventricle with normal dimensions and function. A transesophageal echo at that time confirmed the diagnosis of a membrane-type subaortic stenosis. A further Doppler echo evaluation in January

2002 revealed that the mean transaortic gradient had progressed to 73 mmHg. The patient was reoperated upon in January 2002 and an asymmetric diaphragm was resected. At the immediate and 1-month postoperative follow-up examinations the patient's symptoms and signs (murmur, echo and Doppler findings) were found to have resolved completely. Figure 1 shows the echographic aspects, the catheter pullout curves at cardiac catheterization and the histological images of the resected specimen. While the findings are typical of a membrane-type subaortic stenosis, the case is unusual for its associations, its late presentation and its rapid progression.