

Congenital Sinus of Valsalva Aneurysm with Fistulization into the Right Ventricle Associated with Ventricular Septal Defect

Aneurisma congénito del seno de Valsalva fistulizado al ventrículo derecho asociado con comunicación interventricular

MARTA LÓPEZ RAMÓN, ANA MARCÉN MIRAVETE

These images correspond to a 28 year-old man with history of infundibular ventricular septal defect (VSD) and mild aortic regurgitation secondary to mild prolapse of the right sinus of Valsalva (SV), who underwent VSD closure at the age of 7 with two stitches on teflon and had a remaining residual leak. During follow-up, progressive dyspnea on exertion and continuous murmur over the left sternal border suggestive of fistula were detected. Transesophageal echocardiography showed an 8 mm subaortic, tunneled, infundibular VSD, with drainage into the right ventricular outflow tract (RVOT) (Figure 1), right SV enlargement protruding into the right ventricle (RV) through the VSD with rupture to the RVOT immediately below the pulmonary valve, and mild aortic regurgitation (Figure 2). Cardiac surgery consisting of infundibular VSD closure with a bovine pericardial patch and closure of the right SV perforated to the RVOT was performed, with positive outcome.

Congenital SV aneurysms are the result of dilation in the division between the aortic media and the annulus fibrosus of the aortic valve caused by the action of high blood pressure on a weak point of the wall. When the coronary cusp and the SV wall lack support due to the coexistence of a VSD, the aneurysm penetrates it and protrudes inside the RV causing deformity of the aortic leaflet involved, with secondary valve regurgitation. Finally, the aneurysm may rupture, resulting in a new left-right shunt and congestive heart failure. The frequent association with VSD hampers preoperative diagnostic evaluation. The main limitation of transthoracic echocardiography is the difficult identification of VSD due to flow masking. Transesophageal echocardiography is the most useful diagnostic tool to assess the anatomy and etiopathogenesis of fistulized SV aneurysms, facilitating an optimal approach and surgical treatment.

Conflicts of interest

None declared (See authors' conflicts of interest forms in the website/ Supplementary Material).

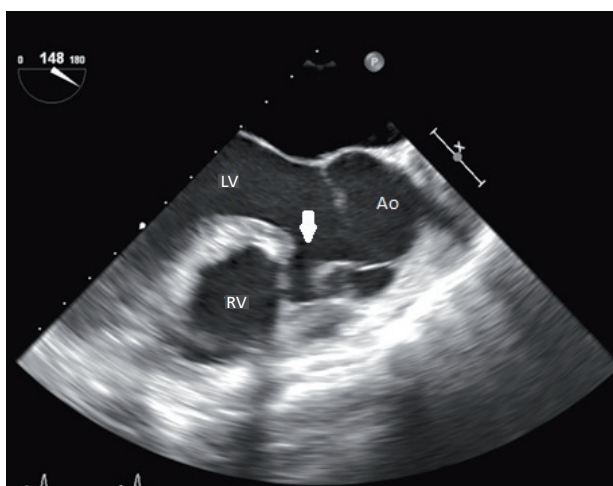


Fig. 1. Transesophageal echocardiogram: subaortic ventricular septal defect (arrow).

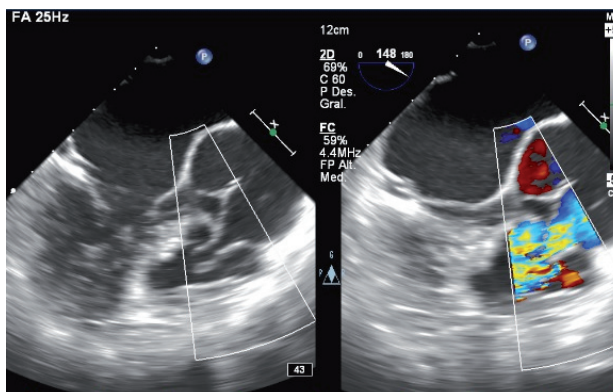


Fig. 2. Transesophageal echocardiogram: 2D image of right Valsalva sinus fistulized to the right ventricle (left) and color Doppler (right) (see color image in the web).