Right ventricular outflow tract obstruction secondary to a membranous ventricular septal aneurysm

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A 55-year-old woman was referred for evaluation of exertional dyspnoea. She was diagnosed in childhood with a bicuspid aortic valve (BAV) and an uncomplicated small perimembranous ventricular septal defect (VSD).

She remained well until the previous year, when she developed progressive NYHA II–III dyspnoea. On TTE, there was no significant stenosis or insufficiency associated with the BAV. A small perimembranous VSD was present with left to right shunting. Colour Doppler interrogation revealed significant flow acceleration in the right ventricular outflow tract (RVOT). TEE confirmed a large (5 × 5 cm) protruding ‘windsock-sock’ type membranous ventricular septal aneurysm (MVSA) obliterating the RVOT. This was associated with prominent right ventricular muscle bundles. The RVOT peak gradient was 91 mmHg. The RV systolic function was normal. There was moderate tricuspid regurgitation. The RV systolic pressure was 120 mmHg (derived from TR jet velocity, assuming right atrial pressure = 10 mmHg; systolic BP was 131 mmHg). The patient underwent repair with resection of the MVSA and the hypertrophied muscle bundles, closure of the small VSD, and annuloplasty of the tricuspid valve. Her functional status improved markedly (NYHA I) following surgery.

MVSA are commonly associated with peri-membranous VSDs and are usually benign. However, in some patients, MVSA can become obstructive and present for the first time in adulthood. Although the most common cause of RVOT obstruction in isolated peri-membranous VSD is the formation of large right ventricular muscle bundles leading to double chamber right ventricular physiology, the presence of an obstructive MVSA should be considered (Figure 1).

Figure 1 (A) Tree-dimensional image of the membranous ventricular septal aneurysm causing right ventricular outflow tract obstruction. (B) Doppler interrogation demonstrating the maximal gradient obtained across the obstruction. (C) Two-dimensional and colour Doppler image of the membranous ventricular septal aneurysm causing right ventricular outflow tract obstruction.