Infective Endocarditis of the Systemic Venous Baffle Following the Atrial Switch Procedure in an Adult

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INTRODUCTION

In dextro-transposition of the great arteries (d-TGA) the aorta arises from a right-sided morphologic right ventricle, and the pulmonary artery arises from a left-sided morphologic left ventricle. For more than two decades, d-TGA was palliated using the atrial switch procedure, whereby an atrial baffle is created from atrial tissue (Senning procedure) or from pericardium or synthetic material (Mustard procedure) to redirect blood flow to the opposite atrioventricular valve and ventricle. In long-term follow-up studies of atrial switch patients, infective endocarditis (IE) is an infrequent cause of morbidity and mortality but has been reported.[1,2] This is the first reported case of IE involving the systemic venous baffle following the atrial switch procedure.

CASE PRESENTATION

A 25-year-old man with a history of d-TGA who had undergone the Senning procedure at 1 week of age was diagnosed with Hodgkin lymphoma 8 months before admission. His initial chemotherapy regimen consisted of chlorambucil/vinblastine/procarbazine/prednisone to limit anthracycline toxicity, given his history of severely reduced systolic function of the systemic right ventricle. He completed two cycles of chlorambucil/vinblastine/procarbazine/prednisone but was believed to be nonadherent to this oral regimen, and 3 months before admission, he was started on the Adriamycin/bleomycin/vinblastine/dacarbazine (ABVD) regimen through a peripherally inserted central catheter (PICC) line. This first cycle of ABVD was complicated by neutropenic fever with coagulase-negative *Staphylococcus* bacteremia. The PICC line was removed, and the patient underwent transesophageal echocardiography (TEE) that showed no evidence of IE. He was treated with antibiotics for a total of 14 days, with resolution of the bacteremia. A new PICC line was placed, and he completed cycle 1 of ABVD. Approximately 1 month later he completed cycle 2 of ABVD but subsequently missed several appointments, and there was concern that he was not receiving adequate PICC care.

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The day before the index hospitalization, the patient presented to the oncology clinic with weakness, dizziness, and hypotension. The patient left the clinic against medical advice but presented to the emergency department the following day, where he was again noted to be hypotensive. Initial vital signs were body temperature 37.5°C, pulse 103 beats/min, blood pressure 81/34 mm Hg, respiratory rate 20 breaths/min, and oxygen saturation 100% on ambient air. Physical examination was notable for somnolence and jaundice, a prominent parasternal heave, a grade II/VI holosystolic murmur along the sternal border, an estimated jugular venous pressure of 8 cm of water, 2+ pedal edema to the thighs, and mild tachypnea. Laboratory studies were notable for the following: white blood cell count 10.0 K/mm^3 (normal 3.8–10.9 K/mm^3), absolute neutrophil count 9.3 K/mm^3 (normal 1.6–8.4 K/mm^3), hemoglobin 8.2 g/dL (normal 13.6–17.3 g/dL), platelet count 94 K/mm^3 (normal 141–401 K/mm^3), sodium 122 mmol/L (normal 136–144 mmol/L), anion gap 14 mmol/L (normal 5–14 mmol/L), blood urea nitrogen 50 mg/dL (normal 8–20 mg/dL), creatinine 3.35 mg/dL (normal 0.5–1.2 mg/dL), estimated glomerular filtration rate 24 mL/min/1.73 m^2 (normal ≥90 mL/min/1.73 m^2), and lactate 2.0 mmol/L (normal 0.5–2.2 mmol/L).

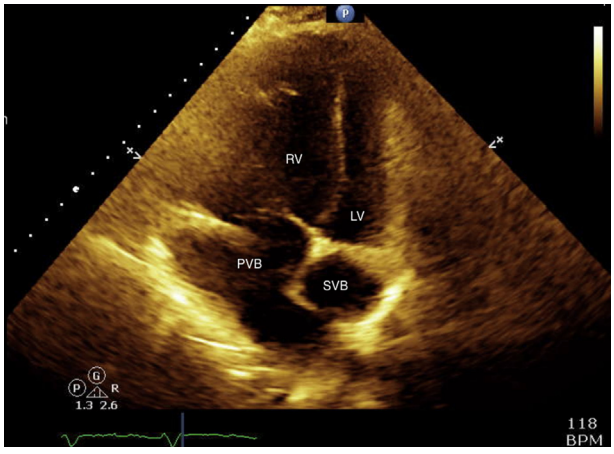

Figure 1 Transthoracic echocardiography, apical four-chamber view, showing the patient's unique anatomy of d-TGA following the atrial switch procedure. The morphologic right ventricle (RV) (systemic ventricle) is severely enlarged and hypertrophied, with bowing of the interventricular septum toward the morphologic left ventricle (LV) (subpulmonic ventricle). The pulmonary venous baffle (PVB) is well seen, whereas the more anterior systemic venous baffle (SVB) is not. There is no evidence of IE in this view.

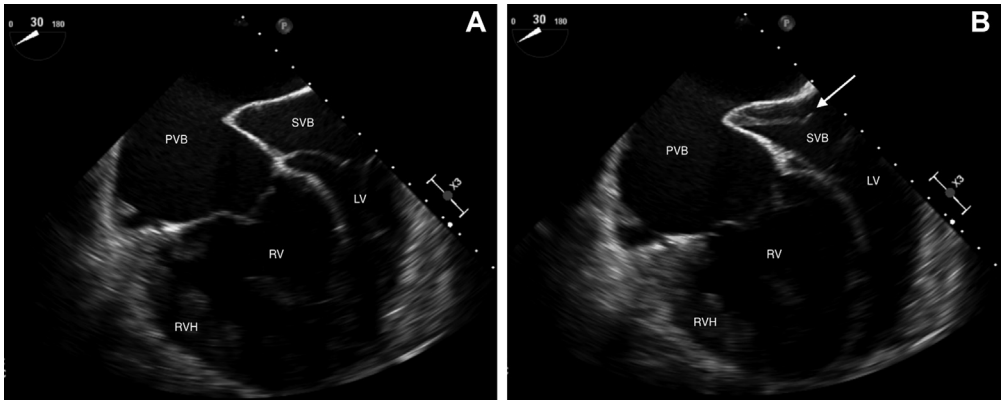

Figure 2 TEE, midesophageal view at 30°, showing a four-chamber view equivalent to the transthoracic view seen in Figure 1. Right ventricular hypertrophy (RVH) is better appreciated on TEE, however. **(A)** No evidence of IE is initially seen. **(B)** After careful angulation toward the more anterior systemic venous baffle (SVB), a large echodensity concerning for IE is noted (*arrow*). *LV*, Left ventricle; *PVB*, pulmonary venous baffle; *RV*, right ventricle.

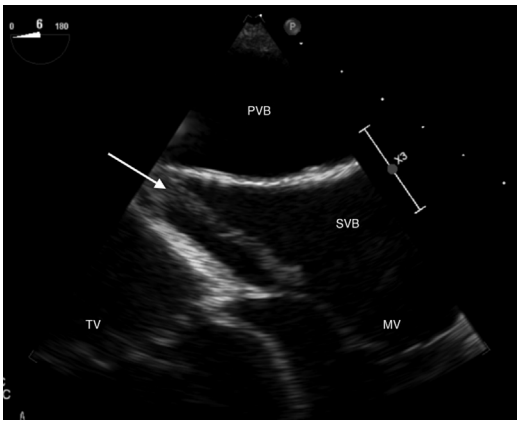

Figure 3 TEE, midesophageal view at 6°, with a zoomed-in view of the large echodensity concerning for IE (*arrow*). *MV*, Mitral valve; *PVB*, pulmonary venous baffle; *SVB*, systemic venous baffle; *TV*, tricuspid valve.

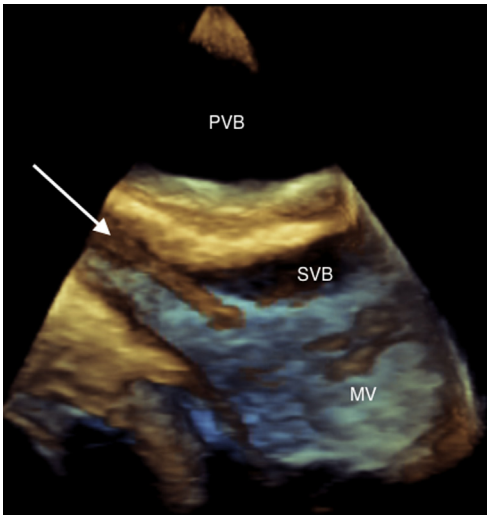

Figure 4 TEE, midesophageal view, with three-dimensional acquisition enabled showing the large echodensity concerning for IE (*arrow*). *MV*, Mitral valve; *PVB*, pulmonary venous baffle; *SVB*, systemic venous baffle.

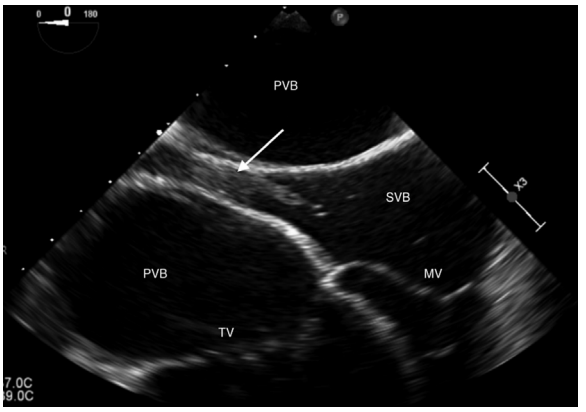

Figure 5 TEE, midesophageal view at 0°, with a zoomed-in view of the large echodensity concerning for IE (*arrow*). The probe has been further advanced from the view in Figure 3 to see the echodensity in its entirety. Note that the echodensity appears to occlude the systemic venous baffle (SVB). *MV*, Mitral valve; *PVB*, pulmonary venous baffle; *TV*, tricuspid valve.

The patient was presumed to be septic and started on empiric broad-spectrum antibiotics following the collection of blood cultures. A central venous catheter was placed, and the PICC line was removed. His blood pressure did not respond to intravenous fluids, and he was started on a norepinephrine infusion. Cardiology was consulted, and transthoracic echocardiography was performed. The results were no different from those of a study completed 2 months prior, with no evidence of IE (Figure 1). Blood cultures were positive for methicillin-sensitive *S. aureus* for two consecutive days. Given this persistent methicillin-sensitive *S. aureus* bacteremia, TEE was performed.

TEE initially showed the same findings as transthoracic echocardiography, with normal systolic function of the morphologic left ventricle (subpulmonic ventricle), severe enlargement and severely reduced systolic function of the morphologic right ventricle (systemic ventricle), and no evidence of IE (Figure 2A). However, careful angulation of the probe toward the more anterior systemic venous baffle revealed a large mobile echodensity in the midportion that initially appeared to be occlusive (Figures 2B–5, Videos 1 and 2). Biplane interrogation of this echodensity showed that it was actually not occlusive but