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



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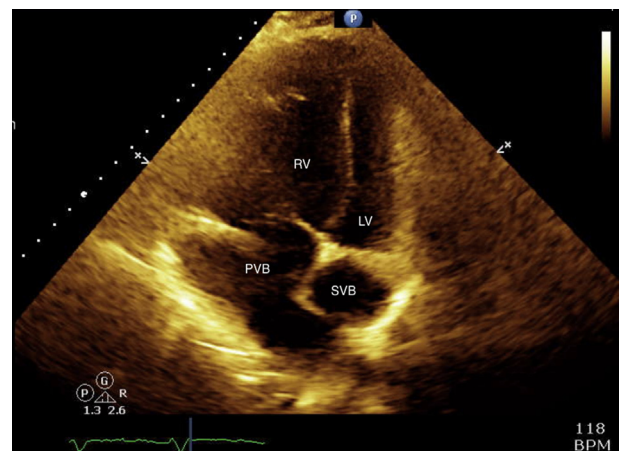
## INTRODUCTION

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.<sup>1,2</sup> 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.

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<sup>3</sup> (normal 3.8–10.9 K/mm<sup>3</sup>), absolute neutrophil count 9.3 K/mm<sup>3</sup> (normal 1.6–8.4 K/mm<sup>3</sup>), hemoglobin 8.2 g/dL (normal 13.6–17.3 g/dL), platelet count 94 K/mm<sup>3</sup> (normal 141–401 K/mm<sup>3</sup>), 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<sup>2</sup> (normal ≥90 mL/min/1.73 m<sup>2</sup>), 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.

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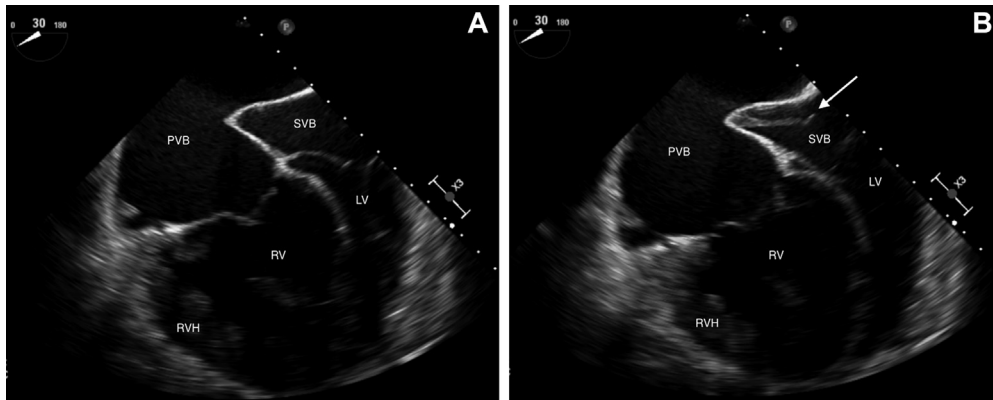
Keywords: Adult congenital heart disease, Echocardiography, Infective endocarditis, Transposition of the great arteries

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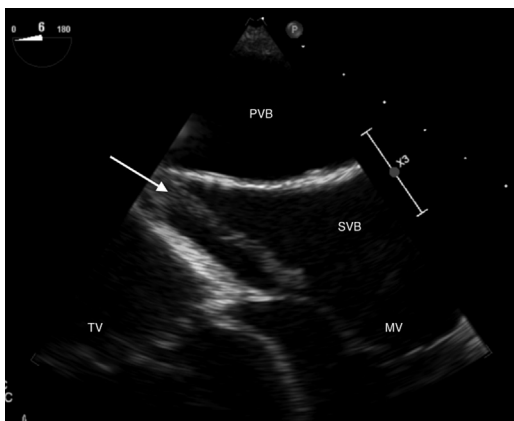
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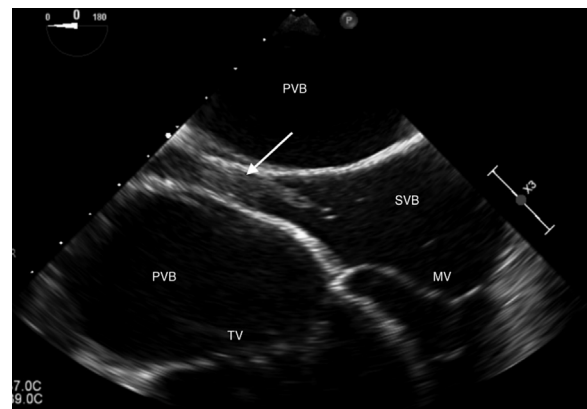
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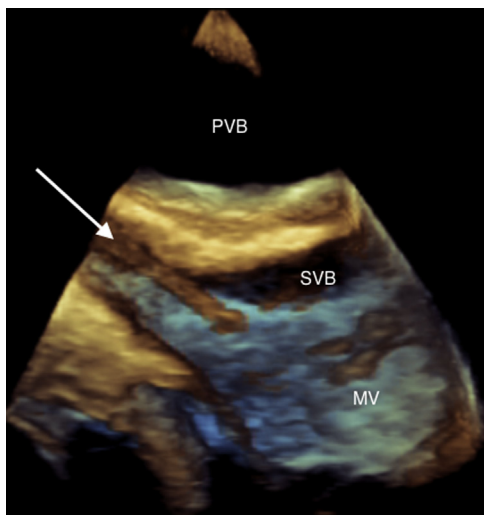
**Figure 2** TEE, midesophageal view at 30°, showing a four-chamber view equivalent to the transthoracic view seen in [Figure 1](#). Right ventricular hypertrophy (RVH) is better appreciated on TEE, however. **(A)** No evidence of IE is initially seen. **(B)** After careful angulation toward the more 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.



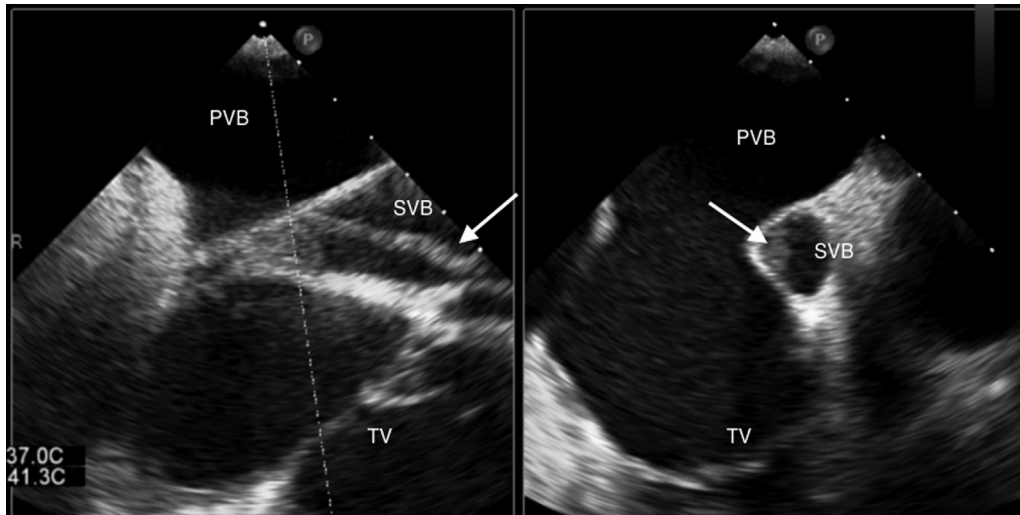
**Figure 5** TEE, midesophageal view at 0°, with a zoomed-in view of the large echodensity concerning for IE (*arrow*). The probe has been further advanced from the view in [Figure 3](#) to see the echodensity in its entirety. Note that the echodensity appears to occlude the systemic venous baffle (SVB).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



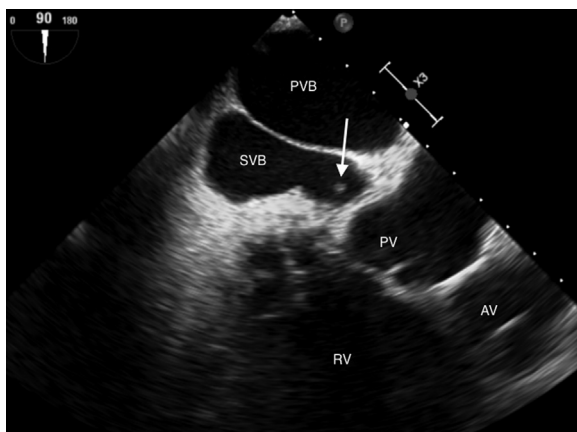
**Figure 4** TEE, midesophageal view, with three-dimensional acquisition enabled showing the large echodensity concerning for IE (*arrow*). MV, Mitral valve; PVB, pulmonary venous baffle; SVB, systemic venous baffle.

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



**Figure 6** TEE, midesophageal view, with biplane imaging focused on the large echodensity concerning for IE (arrow). The four-chamber view (left) suggests that the echodensity may be occluding the systemic venous baffle (SVB), but the orthogonal view (right) shows that the echodensity (arrow) is eccentric in the SVB and primarily adherent to the posterior surface. PVB, Pulmonary venous baffle; TV, tricuspid valve.



**Figure 7** TEE, midesophageal view at 90°, showing the side-by-side orientation of the great arteries resembling a “double-barrel shotgun.” Note that the aortic valve (AV) is more anterior and rightward of the pulmonic valve (PV) and arises from the morphologic right ventricle (RV) (systemic ventricle), consistent with d-TGA. The echodensity concerning for IE is incidentally noted en face (arrow) in the systemic venous baffle (SVB). PVB, Pulmonary venous baffle.

eccentric (Figure 6). Other characteristic anatomy and sequelae of d-TGA were demonstrated on TEE, including the parallel orientation of the great arteries resembling a “double-barrel shotgun” (Figure 7),<sup>3</sup> subpulmonic stenosis due to bulging of the interventricular septum toward the morphologic left ventricle (subpulmonic ventricle) (Figure 8), and significant tricuspid regurgitation (systemic atrioventricular valve) (Figure 9). At the conclusion of the study, an agitated saline injection (“bubble study”) was performed and excluded any significant baffle leaks (Figure 10).

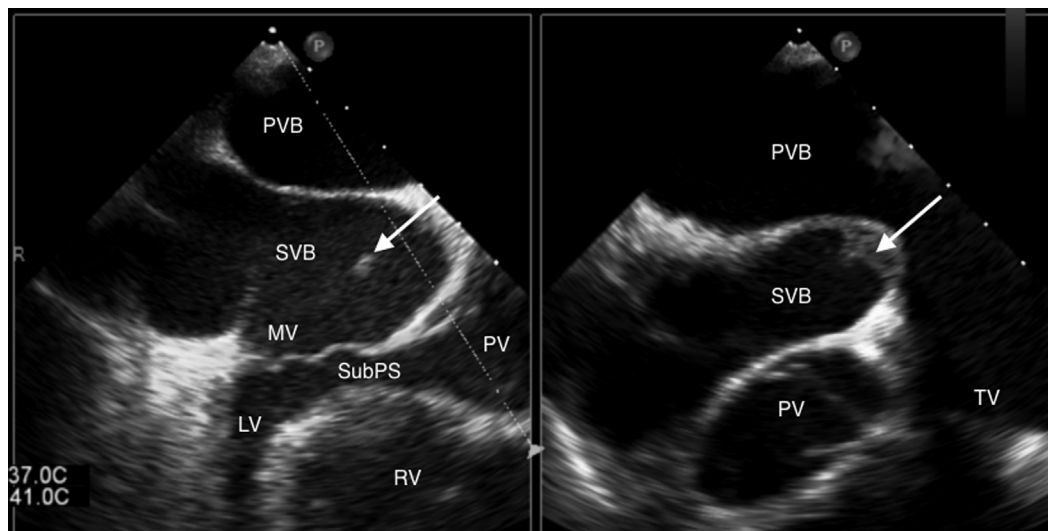
Given concern for a superinfected thrombus, the patient was started on anticoagulation in addition to continuing antibiotics.

The patient failed to improve clinically, with worsening renal function requiring dialysis, worsening liver function, encephalopathy, persistent fevers, and a persistent pressor requirement. Repeat TEE was performed 1 week later and showed no improvement in the size of the vegetation. The patient was deemed too ill for surgical debridement of the systemic venous baffle, and the decision was made to pursue percutaneous aspiration with an AngioVac cannula (AngioDynamics, Latham, NY).

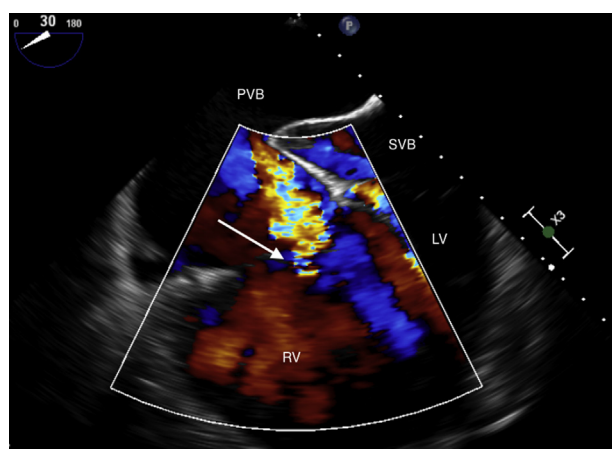
The day before the AngioVac procedure, the patient was electively intubated for hemodynamic stabilization, but he developed bradycardia and pulseless electrical activity. Cardiopulmonary resuscitation was continued for nearly 2 hours, but the patient ultimately expired. Autopsy revealed a 1 × 1 cm thrombus in the superior vena cava, a 4 × 3 × 1 cm thrombus in the systemic venous baffle, a 1 × 1 cm pulmonary embolus, and a vegetation on the left-sided atrioventricular (mitral) valve (this was not visualized on TEE). Multiple scattered nodules were also found in the liver and lungs. On microscopic examination, the systemic venous baffle thrombus was noted to have acute inflammation, fibrin deposition, and hemorrhage. Additionally, active Hodgkin lymphoma involvement was found throughout the reticuloendothelial system. The final cause of death by autopsy was not definitive.

## DISCUSSION

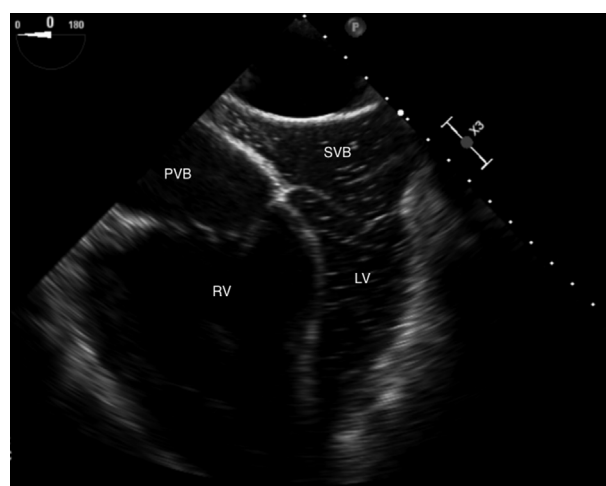
Although the atrial switch is no longer performed routinely for the palliation of d-TGA, many adults have had this procedure and are at risk for its sequelae. Additionally, the double switch operation for levo-transposition of the great arteries will ensure that the atrial switch continues to be encountered. Well-known sequelae include arrhythmias, reduced systolic function of the systemic (morphologic right) ventricle, tricuspid regurgitation, and baffle leaks and/or obstruction.<sup>4</sup> The superior limb and midportion of the systemic venous baffle are particularly prone to stenosis, and it was likely the combination of this condition and the PICC line that precipitated thrombus formation in the patient presented here.



**Figure 8** TEE, midesophageal view, with biplane imaging focused on the large echodensity concerning for IE (arrows). Subpulmonic stenosis (SubPS) is noted due to the bulging of the interventricular septum toward the lower-pressure morphologic left ventricle (LV) (subpulmonic ventricle). MV, Mitral valve; PV, pulmonic valve; PVB, pulmonary venous baffle; RV, right ventricle; SVB, systemic venous baffle; TV, tricuspid valve.



**Figure 9** TEE, midesophageal view at 30°, with color flow Doppler imaging applied showing significant tricuspid regurgitation (arrow). LV, Left ventricle; PVB, pulmonary venous baffle; RV, right ventricle; SVB, systemic venous baffle.



**Figure 10** TEE, midesophageal view at 0°, following the injection of agitated saline (“bubble study”). Note that the systemic venous baffle (SVB) and morphologic left ventricle (LV) (subpulmonic ventricle) are filled with bubbles, while the pulmonic venous baffle (PVB) and morphologic right ventricle (RV) (systemic ventricle) are not, indicating no evidence of a significant baffle leak.

Clinicians caring for atrial switch patients should exclude baffle stenosis before placing PICC lines, as is the standard of care before placing transvenous pacemakers. It is possible for patients to develop baffle stenosis and obstruction without overt symptoms of superior vena cava syndrome because of collateral flow via the azygos system.<sup>5</sup> It is similarly important to exclude significant baffle leaks with an echocardiographic “bubble study,” as was done in this case, because the presence of a leak can predispose to paradoxical emboli (or a brain abscess in the setting of a vegetation) with devastating consequences. For these reasons, consultation with an expert in the field of adult congenital heart disease (ACHD) is highly recommended. One consideration for patients at higher risk for IE is a venous port, which has a markedly lower risk for central line–associated bloodstream infection compared with PICCs.<sup>6</sup>

The definitive diagnosis of IE was based on the modified Duke criteria, wherein the patient described here fulfilled two major criteria: typical microorganisms from two separate blood cultures and TEE showing a mobile intracardiac mass suspicious for a vegetation.<sup>7</sup> Although the patient had a history of methamphetamine use, he had no history of intravenous drug use, and the methicillin-sensitive *S. aureus* bacteremia in this case was most likely the result of poor PICC care. Management of “right-sided” IE is usually conservative, but evidence of persistent infection by serial TEE and a poor clinical response to antibiotic therapy prompted consideration for surgical

debridement. Multiple organ failure and hemodynamic instability made him a poor surgical candidate, however. Percutaneous aspiration of a right-sided vegetation has been reported previously with excellent results, and this was believed to be the best option for this particular patient.<sup>8</sup>

Despite IE occurring at a rate 10-fold higher in patients with ACHD, there is a relative paucity of data on the topic. A recent review found just over 20 studies in total examining IE in patients with ACHD, with the majority of the data being 15 years old.<sup>9</sup> In one small ACHD database ( $N = 185$ ), there were no cases of IE in patients with the atrial switch procedure,<sup>10</sup> but a much larger Dutch registry ( $N = 10,210$ ) reported that 3% of those with IE had d-TGA as the underlying defect (type of repair not specified).<sup>11</sup> In general, the risk for endocarditis following the atrial switch procedure is considered low, though certain residua (such as ventricular septal defects) and sequelae (subpulmonic stenosis and tricuspid regurgitation) may increase this risk. Nonetheless, published guidelines regarding the prevention of endocarditis generally advise against prophylaxis in this patient population. Still, some authors argue that prophylaxis is warranted given the presence of a high-velocity jet (tricuspid regurgitation) near prosthetic material (atrial baffle).<sup>12</sup>

## CONCLUSION

Descriptions of IE or thrombosis involving shunts or baffles in repaired congenital heart disease are extremely limited in the medical literature. Successful AngioVac thrombectomy has previously been described in a patient with Fontan circulation.<sup>13</sup> To our knowledge, this is the first report of a superinfected thrombus involving the systemic venous baffle in a patient following the atrial switch procedure. TEE with careful attention to the patient's unique cardiac anatomy was essential to the diagnosis of IE in this case.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.case.2017.02.005>.

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