A 43-year-old woman presented with shortness of breath. She developed dyspnea on exertion 6 months prior to presentation, with resting shortness of breath for 2 months. Dyspnea was worse in the upright position. She recently noted that her lips were turning blue in color. Her past medical history was significant for intravenous drug abuse, hepatitis C, and tricuspid valve (TV) endocarditis 3 years prior. She underwent TV replacement with a bioprosthetic valve that was complicated by chest wall necrotizing fasciitis and septic embolism with brain abscess requiring craniotomy. She reported tobacco use, medication noncompliance, and continued intravenous drug abuse. Medications included albuterol/ipratropium, pantoprazole, and furosemide. She had been started on continuous home oxygen. She presented to another institution and was found to have a patent foramen ovale (PFO). Coronary angiography showed no significant coronary artery disease, and chest computed tomography angiography demonstrated normal lung parenchyma and no pulmonary embolus. She was referred to our institution for percutaneous PFO closure.

Admission vital signs and basic labs are shown in Table 1. Pulse oximetry on 5L continuous oxygen by nasal cannula was 75% to 78%. When upright, her oxygen saturation decreased to 65% and she became more tachypneic, findings consistent with the platypnea-orthodeoxia syndrome. Physical examination showed perioral cyanosis, no jugular venous distention, clear lungs, and no cardiac murmurs. Arterial blood gas on 40% FiO2 demonstrated a chronic respiratory alkalosis with a compensatory metabolic acidosis. The electrocardiogram showed sinus tachycardia. An echocardiogram showed sinus tachycardia. An echocardiogram demonstrated a 9 mm Hg gradient across the TV, which was otherwise not well seen (Movie 1, Figure 1A). Right heart catheterization confirmed a 9 mm Hg gradient, consistent with severe TV stenosis (Figure 1B), and an oximetry run demonstrated arterial desaturation without left-to-right shunt (Table 2). Pulmonary angiography did not show arteriovenous malformations (Movie 2). Inferior vena cava venography demonstrated simultaneous opacification of the right ventricle and left atrium, consistent with a large right-to-left interatrial shunt (Movie 3). Transesophageal echocardiography showed significant thickening of the bioprosthetic TV leaflets with severely restricted leaflet motion and a large mass of low echo density (Movies 4A and 4B). 3D imaging showed a severe reduction in valve orifice area with a slit-like appearance (Movie 5). The transesophageal echocardiography also confirmed a significant right-to-left interatrial shunt through a large PFO (Figure 2, Movie 6). Cardiac magnetic resonance imaging confirmed the shunt (Figure 3, Movie 7), showed the TV mass (Figure 4, Movie 8) and demonstrated a Qp/Qs of 0.5.

The patient was started on anticoagulation and high-flow oxygen but remained markedly hypoxemic and symptomatic due to the shunt. Because of the large PFO and possibility of embolism, fibrinolytics were not considered. Early valve failure may have been due to noncompliance with warfarin treatment after the initial operation or the inability to treat with warfarin due to perioperative complications at that time. A hypercoagulability work-up was negative. There was no
other evidence for infection to suggest subacute bacterial endocarditis.

The patient suffered a stroke; brain MRI findings suggested paradoxical embolism through the PFO. She had good neurological recovery. Because of the possibility of recurrent stroke (which contraindicates fibrinolytics) and the severity of symptoms related to the shunt, it was decided that surgical intervention was necessary, despite high operative risk. TV valvulectomy without replacement has been used in the past, primarily for TV endocarditis. However, it is complicated by ubiquitous elevation of right-sided pressures, overt right heart failure in at least 30%,1,2 and it often requires reoperation for valve placement.3 Given her history of noncompliance, a mechanical valve was not considered. She underwent successful TV replacement with a bioprosthetic TV and intraoperative PFO closure (Figure 5, Movies 9 and 10). All symptoms resolved, and she was discharged in good condition. Pathology showed fibrous exudates on the TV consistent with organized thrombus; Gram’s stain and cultures were negative.

Atrioventricular valve stenosis can be associated with interatrial shunting due to elevated atrial pressures. Lutembacher syndrome, originally described in 1916, occurs when

Table 2. Oxymetry Saturation Run Performed During Right Heart Catheterization

<table>
<thead>
<tr>
<th>Location</th>
<th>Saturation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>53%</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>60%</td>
</tr>
<tr>
<td>High right atrium</td>
<td>57%</td>
</tr>
<tr>
<td>Low right atrium</td>
<td>58%</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>58%</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>56%</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>84%</td>
</tr>
</tbody>
</table>

Test performed with patient on 40% FiO2.
an interatrial left-to-right shunt develops due to elevated left atrial pressure secondary to mitral stenosis. The triad of TV stenosis, elevated right atrial pressure, and right-to-left shunt has been called reverse Lutembacher syndrome. There have been two reported cases of this syndrome. Both cases occurred in patients with rheumatic valvular disease presenting with progressive shortness of breath and hypoxemia. The first patient underwent transseptal mitral and aortic valvuloplasty 4 years prior to presentation, and later developed rheumatic TV stenosis with right-to-left shunting through the iatrogenic atrial septal defect. The second patient presented with isolated TV stenosis and shunting through a PFO. The mechanism for the platypnea-orthodeoxia syndrome is not clearly understood, but altered streaming of blood in the right atrium while upright may cause shunting across a PFO, particularly in states of elevated right atrial pressure.

This is the first description of reverse Lutembacher syndrome secondary to bioprosthetic TV stenosis. It is also the first reported case of paradoxical embolism with reverse Lutembacher physiology, and is the first description of the platypnea-orthodeoxia syndrome associated with reverse Lutembacher syndrome or TV stenosis. Imaging was central to making the correct diagnosis and choosing the proper management strategy.

Disclosures

None.

References

Platypnea-Orthodeoxia Syndrome Associated with Bioprosthetic Tricuspid Valve Stenosis and Reverse Lutembacher Syndrome
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Circulation. 2011;123:e222-e224
doi: 10.1161/CIRCULATIONAHA.110.979427
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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