Amelioration of High Cardiac Output and Pulmonary Hypertension by Occlusion of Congenital Porto-Systemic Shunt

Paul M. McKie, MD; Robert B. McCully, MD; Patrick S. Kamath, MD; Thomas C. Bower, MD; Michael A. McKusick, MD; Andre C. Lapyere, MD; Naveen L. Pereira, MD

A 69-year-old woman was evaluated for progressive dyspnea and pulmonary hypertension. She had World Health Organization class III symptoms, was obese (41 kg/m² body mass index), and had chronic atrial fibrillation, obstructive sleep apnea, and mild chronic obstructive pulmonary disease. Positive findings on physical examination included an elevated jugular venous pressure (14 cm), irregular rhythm, accentuated pulmonic second sound, and lower extremity edema. Blood pressure was 122/60 mm Hg, and heart rate was 68 beats per minute. The distance covered during a 6-minute walk test was 266 m. Transthoracic echocardiogram documented preserved left ventricular ejection fraction, moderate right ventricular enlargement, mild decrease in right ventricular function, and estimated right ventricular systolic pressure of 102 mm Hg, assuming a right atrial pressure of 10 mm Hg (Table 1 and online-only Data Supplement Movie 1). Severe tricuspid regurgitation and trivial mitral regurgitation were present without other valvular abnormalities. Aortic flow was elevated, and the calculated cardiac index was 4.2 L/min/m². Right heart catheterization demonstrated a right atrial pressure of 11 mm Hg, mean pulmonary artery pressure of 46 mm Hg, mean pulmonary capillary wedge pressure of 19 mm Hg, and cardiac output (CO) of 13.4 L/min (6.8 L/min/m² cardiac index, Fick method), consistent with pulmonary hypertension and high output heart failure. Oxygen saturation studies, performed as part of a shunt run, demonstrated the following: superior vena cava 74%, mid-right atrium 79%, and inferior vena cava 87%, suggestive of a systemic shunt. Thyroid function (thyroid stimulating hormone, 1.9 mIU/L) was normal, and hemoglobin was 12.7 g/dL. Computed tomography of the chest, abdomen, and pelvis did not demonstrate an arteriovenous fistula, but there was evidence of a large congenital venous malformation with a shunt between the inferior mesenteric vein and inferior vena cava (Figure). The porto-systemic shunt was confirmed on mesenteric angiogram/venogram with preferential blood flow from the splenic vein through the inferior mesenteric vein and then into the inferior vena cava via a tangle of collateral vessels in the retroperitoneum. The net effect of this pathway resulted in shunting of most of the splenic venous blood flow back to the systemic circulation, bypassing the liver.

The porto-systemic shunt was embolized with coils and Amplatzer plugs (Figure, C and D) after plasma sampling at

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<tr>
<th>Table 1. Catheterization and Echocardiographic Hemodynamic Assessment Before and After Closure of the Congenital Anomalous Shunt</th>
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<td><strong>Before</strong></td>
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<tr>
<td><strong>Catheterization derived</strong></td>
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<tr>
<td>Cardiac output (Fick method), L/min</td>
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<tr>
<td>Right atrial pressure, mean, mm Hg</td>
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<td>Right ventricle end diastolic pressure, mm Hg</td>
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<td>Pulmonary artery pressure, mean, mm Hg</td>
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<td>Pulmonary capillary wedge pressure, mean, mm Hg</td>
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<td><strong>Echocardiographic derived</strong></td>
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<tr>
<td>Right ventricle internal dimension, mid-cavity,* mm</td>
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<tr>
<td>Right ventricular systolic pressure, mm Hg</td>
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<tr>
<td>Tricuspid annular plane systolic excursion, mm</td>
</tr>
<tr>
<td>Right ventricular index of myocardial performance, no units</td>
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<tr>
<td>Right ventricular strain, average, %</td>
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<tr>
<td>Left ventricular end diastolic diameter, mm</td>
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<tr>
<td>Left ventricular end systolic diameter, mm</td>
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<tr>
<td>Left atrial volume index, mL/m²</td>
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<tr>
<td>Right atrial volume index†, mL/m²</td>
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*Measurement at end diastole.
†Patient weight decreased 11 kg after shunt closure.

From the Divisions of Cardiovascular Diseases (P.M.M., R.B.M., A.C.L., N.L.P.), Gastroenterology (P.S.K.), Vascular Surgery (T.C.B.), and Vascular and Interventional Radiology (M.A.M.), Mayo Clinic and Foundation, Rochester, MN.

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multiple levels (Table 2). Six months after embolization, the patient improved symptomatically with only World Health Organization class I–II symptoms. The 6-minute walk test distance increased by 71 m. Echocardiogram demonstrated lower estimated right ventricular systolic pressure, moderate tricuspid regurgitation, and improved right ventricular parameters with reduced right ventricular size and normalized right ventricular systolic function (Table 1 and online-only Data Supplement Movie II). Right heart catheterization demonstrated a marked decrease in CO and improvement in mean pulmonary artery pressure. A complete oxygen saturation run demonstrated no residual intra-abdominal shunt. On most recent follow-up 1 year after embolization, the patient continues to improve symptomatically.

Ohm’s law suggests that pulmonary hypertension is secondary to increased pulmonary resistance or increased pulmonary flow. The most common causes of high CO and increased pulmonary flow include anemia, renal disease, cirrhosis, hyperthyroidism, arteriovenous fistulae, and intracardiac shunts, which were all absent in this case. We have described previously the association between spontaneous porto-systemic shunts in patients with cirrhosis and porto-pulmonary hypertension. This patient had high CO as a result of a congenital porto-systemic shunt in the absence of cirrhosis. The congenital porto-systemic shunt was likely the cause of the pulmonary hypertension because occlusion of the shunt resulted in amelioration of symptoms with reduction in CO and mean pulmonary artery pressure. The lack of demonstration of a gradient in endothelin or atrial natriuretic peptide (Table 2) across the shunt confirms that the pulmonary hypertension was not related to alteration of these vasoactive substances from the splanchnic circulation. In conclusion, this case highlights congenital porto-systemic venous shunts as an unusual and potentially overlooked cause of high CO and pulmonary hypertension, treatment of which can result in symptomatic and functional improvement.

Acknowledgments
We thank Drs John Burnett and Lilach Lerman for measurement of atrial natriuretic peptide and endothelin levels.

Disclosures
None.

References

Table 2. Endothelin and Atrial Natriuretic Peptides Before and After Closure of the Congenital Anomalous Shunt

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<tr>
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<th>Before Closure</th>
<th>After Closure</th>
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<tr>
<td></td>
<td>Portal Vein</td>
<td>Below Shunt</td>
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<tr>
<td>Atrial natriuretic peptide, pg/mL</td>
<td>74.7</td>
<td>73.9</td>
</tr>
<tr>
<td>Endothelin, pg/mL</td>
<td>35.6</td>
<td>37.0</td>
</tr>
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Figure. Venography (top) and computerized axial tomography scan (bottom) images before (A and B) and after (C and D) closure of the congenital venous malformation (thin arrow), which formed a low-pressure/high-flow anomalous venous communication (arrowhead) from the inferior mesenteric vein to inferior vena cava. Thick arrow, Amplatzer plugs; thin arrow, venous malformation; double thin arrow, embolization coils; arrowhead, communication from inferior mesenteric vein to inferior vena cava. IVC indicates inferior vena cava; IMV, inferior mesenteric vein; PV, portal vein; SV, splenic vein; and Ao, aorta.
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