A 7-month-old boy presented with significant cyanosis 17 days after bidirectional cavopulmonary shunting (Glenn procedure). He had previously undergone a Norwood operation at 7 days of age for a double-outlet right ventricle with hypoplastic left ventricle, hypoplastic aortic arch, and coarctation. Cardiac catheterization 10 days before the Glenn operation showed a pulmonary venous wedge pressure of 16 mm Hg and an estimated pulmonary vascular resistance of 3 U·m². Angiography demonstrated moderate right pulmonary artery stenosis and a 0.6 mm vein draining from the left end of the innominate vein into the posterior mediastinum (Figure, a).

The patient underwent a bidirectional Glenn shunting with placement of a right pulmonary artery patch. He was discharged home on postoperative day 8, with a systemic saturation of 82% on room air. At follow-up on postoperative day 17, he was in mild respiratory distress with a saturation of 60% on room air. Chest x-ray showed clear lung fields bilaterally. Contrast echocardiography with agitated saline injected in a scalp vein demonstrated rapid contrast return to the left atrium. A pulmonary perfusion scan with tracer injected in a scalp vein showed a right-to-left shunt with 60% of tracer passing to the body and 40% to the lungs.

Cardiac catheterization on the following day showed a superior caval pressure of 20 mm Hg and a systemic oxygen saturation of 73% (ventilated with 50% oxygen). Angiography showed a large, 6.4-mm venous fistula from the left end of the innominate vein to the left upper pulmonary vein (Figure, b). Through a shortened 5F delivery sheath placed in the left internal jugular vein, an 8/6 mm Amplatzer duct occluder was deployed in the distal fistula. A subsequent angiogram demonstrated complete occlusion of the vessel (Figure, c). Systemic oxygen saturation immediately improved to 83%, without an increase in superior vena cava pressure.

This case illustrates the rapid development of a large decompressing venous fistula within 17 days of bidirectional Glenn shunt. Contributing factors likely included the presence of a substrate vessel (a tiny vestigial levoatriocardinal vein) and high superior vena cava pressure.
A. Left innominate vein angiogram in posteroanterior projection, 10 days before bidirectional Glenn shunting, demonstrating a tiny venous connection to the posterior mediastinum. B. Left internal jugular vein angiogram in posteroanterior projection, 18 days after bidirectional Glenn shunting, demonstrating a large 6.4-mm venous fistula to the left upper pulmonary vein. C. Left internal jugular vein angiogram 10 minutes after placement of the Amplatzer duct occluder, demonstrating complete occlusion.
Rapid Development of a Large Systemic-to-Pulmonary Vein Fistula After Bidirectional Glenn Shunt and Successful Closure With an Amplatzer Duct Occluder
Muhammad Dilawar, William M. Gottliebson, Scott M. Bradley and Wolfgang A.K. Radtke

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