Very Large Patent Ductus Arteriosus With Severe Pulmonary Arterial Hypertension

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A 34-year-old woman was referred due to a patent ductus arteriosus (PDA) and pulmonary hypertension (PH). She presented with oxygen saturation 92–93% in both the upper and lower limbs. Transthoracic echocardiography indicated significant PDA flow (Figure A). Computed tomography showed a very large elongated-type PDA (Figure B,C). On cardiac catheterization, mean pulmonary arterial pressure (mPAP) was 74 mmHg; pulmonary-to-systemic pressure (Pp/Ps), 0.85; and pulmonary vascular resistance (PVR), 14.0 Wood units; whereas left-to-right shunt (Qp/Qs; 1.75) was still prevalent. Aortography confirmed that the contrast medium drained toward the pulmonary trunk (Figure D; Supplementary Movie 1). We decided to trial PDA occlusion to clarify PH reversibility (Figure E), which markedly decreased mPAP (41 mmHg) and Pp/Ps (0.42) to below 0.5, the cut-off for persistent PH.\(^1\) Nitric oxide inhalation further improved mPAP (33 mmHg) during occlusion. Because the PDA was too large for the PDA occluders, we deployed a 24-mm Amplatzer Septal Occluder®, which had a waist diameter 71% larger than the pulmonary end,\(^2\) without developing a pressure gradient inside the aorta or the pulmonary artery, after institution ethics committee approval (Figure F). The moderate shunt remaining after the procedure had disappeared almost completely 6 months later (Figure G; Supplementary Movie 2), with improved mPAP (39 mmHg), PVR (11.0 Wood units), and oxygen saturation (97–100%). We introduced 10-mg/day macitentan, an endothelin receptor antagonist. This repair-and-treat strategy consequently improved mPAP and PVR to 29 mmHg and 8.0 Wood units, respectively.

Disclosures
The authors declare no conflicts of interest.

References

Supplementary Files
Supplementary Movie 1. Descending aortography, a lateral projection showing that contrast medium drains from the aorta mostly in the direction of the pulmonary trunk.
Supplementary Movie 2. At 6 months after the successful device closure of the patent ductus arteriosus, descending aortography, a lateral projection showed that the residual shunt had disappeared almost completely.

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Figure. (A) Transthoracic echocardiography showing patent ductus arteriosus (PDA) flow toward the pulmonary valve (arrow). (B, C) Computed tomography demonstrating the very large PDA (arrow); the ampulla (39 mm) and the pulmonary end (14 mm) (asterisks). (D, E) Aortography confirming (D) a type A Krichenko PDA (conical duct with prominent aortic ampulla and constriction near the pulmonary end) and (E) complete occlusion during trial occlusion. (F, G) Fluoroscopy showing a 24-mm Amplatzer Septal Occluder® (F) immediately and (G) 6 months after the implantation, which continued to close during the 6 months. PA, pulmonary artery.