The surgical treatment of drug-refractory idiopathic pulmonary arterial hypertension at the level of the interatrial septum has been the choice of treatment until the emergence of the reversed Potts shunt (rPS). Since its introduction, rPS has been used in an increasing frequency, even as an alternative to lung or heart-lung transplantation in symptomatic patients with suprasystemic pulmonary arterial pressures to mimic Eisenmenger’s physiology beyond supra-aortic vessels. Although confined to small cohorts, initial results with this approach have been promising, with prompt improvement in right ventricular systolic function, quality of life, and prolonged survival; however, the indication and the optimal technical approach remain obscure. The latest alteration to the classic rPS was the creation of a unidirectional valved shunt (UVS), which unloads the right ventricle and prevents bidirectional shunting in early systole as the result of delayed right ventricular contraction or ventricular desynchronization that is observed in pulmonary hypertension. Surgically, this was performed by sewing a bovine jugular vein graft into a standard polytetrafluoroethylene graft. Herein, we present a novel UVS design with a 10-mm femoral vein graft in a 10-mm Gore-Tex graft in a symptomatic patient with refractory pulmonary hypertension and chronic lung disease.

METHODS
A waiver of documented consent was granted by the Children’s National Hospital Institutional Review Board. The patient was a 12-year-old girl with pulmonary hypertension associated with a moderate-sized atrial septal defect, history of low birth weight (500 g), prematurity (24 weeks), and chronic lung disease. At 1 year of age, she had been admitted to the hospital due to respiratory syncytial virus infection, which had necessitated venovenous extracorporeal membrane oxygenation. She presented to our hospital with markedly reduced exercise capacity (New York Heart Association functional class III) and asthma-like symptoms despite triple therapy with selective IP prostacyclin-receptor agonist, phosphodiesterase-5 inhibitor, and endothelin receptor antagonist.

Surgery was performed via median sternotomy. Cardiopulmonary bypass was established through routine aorta-bicaval cannulation, and the patient was cooled down to 28 °C. Following aortic crossclamping and antegrade Custodiol cardioplegia, a large ostium secundum atrial septal defect was closed through right atriotomy with a CardioCel neo patch (LeMaitre Vascular) using a continuous 5-0 PROLENE suture (Ethicon). A 4-mm fenestration was performed on the patch, through which the left heart was deaired. After the removal of the crossclamp, a valved rPS that was already prepared using a 10-mm femoral vein homograft with a valved section was inserted into a 10-mm Gore-Tex graft (Video 1). After measurement of the length, the vein graft was sutured on both ends to the Gore-Tex graft using 7-0 PROLENE in a continuous fashion. A few fenestrations were created on the Gore-Tex graft to prevent compression due to possible bleeding between the homograft and Gore-Tex tube. On cardiopulmonary bypass, the descending aorta and the isthmus region were side clamped, with continuous near-infrared spectroscopy monitoring of the
lower body. Following an appropriate incision, the Potts shunt graft was sutured to the descending aorta distal to the subclavian artery using a 6-0 C1 suture. Then, the proximal part was attached to the main pulmonary artery using a 6-0 PROLENE suture. Postoperative transesophageal echocardiogram revealed a good left ventricular function, moderately depressed right ventricular function similar to the preoperative level, and right-to-left blood flow through the Potts shunt. A delayed chest closure was performed later. The patient was discharged home on the 13th postoperative day and was doing well on her last follow-up at 3 months.

DISCUSSION
In patients with refractory pulmonary hypertension with infra- or systemic pulmonary arterial pressures and exercise intolerance, placement and bidirectional flow through the rPS could potentially worsen right ventricular function and pulmonary hypertension. Initial experiences with UVS with flap-valves or jugular vein homografts have been promising. Considering the increased infective endocarditis risk of bovine jugular venous valved conduits and the potential need for anticoagulation after rPS surgery, we preferred using a reinforced cryopreserved valved femoral vein graft as an alternative. We have previously published our experience with this graft in the right ventricular outflow tract position, which has yielded comparable results with aortic or pulmonary homografts. Therefore, we suggest the consideration of reinforced femoral vein grafts for the creation of rPS, although longer follow-up with a larger group of patients is warranted to determine the durability of this conduit and the prognosis of the patients.

CONCLUSIONS
Unidirectional rPS can alleviate the worsening of the right ventricular function and pulmonary circulation due to the intermittent bidirectional flow through the aortopulmonary shunt in patients with pulmonary hypertension with infra- or systemic pulmonary artery pressures. In addition to the previous rPS designs with flap valves or jugular vein homografts, a femoral vein homograft could be a viable alternative and lessen the risk of infective endocarditis and need for anticoagulation. When reinforced with a PTFE conduit, sustained valve integrity under systemic pressures can be expected.

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References