

# Combined strategy of Waterston shunt percutaneous occlusion and medical treatment with sildenafil for management of pulmonary hypertension in an adult patient with corrected tetralogy of Fallot

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A female patient with diagnosis of severe tetralogy of Fallot (TOF) underwent a palliative Waterston shunt (WS) at 5 months of age [1]. Three years later, corrective surgery was performed in another hospital. The surgical report stated closure of the WS. The patient remained in NYHA II and had two successful pregnancies. At the age of 38 years, she was admitted to our hospital for worsening of symptoms (NYHA III). The computed tomography (CT) angiogram showed patency of the WS (Figures 1 A and B). At the right heart catheterization, the pressures before and after the nitric oxide test were 79/39/54 mm Hg and 73/35/48 mm Hg in the pulmonary artery (PA), and 125/66/89 and 140/65/94 mm Hg in the aorta (Ao), respectively. Moreover, the pressure gradient of the right pulmonary artery (RPA) close to the WS was 16 mm Hg, whereas the proximal and distal PA pressures to the shunt were 79/39/54 and 63/36/49, respectively. Pulmonary vascular resistance (PVR) was 2.2 Wood units (WU), and pulmonary to systemic flow ratio calculated by the Fick formula ( $Q_p/Q_s$ ) was 3.1. Moreover, the occlusion test with a TyShak balloon was positive. The patient was treated with 75 mg of sildenafil [2] for 6 months, obtaining a PA pressure of 78/37/52 mm Hg, an Ao pressure of 125/66/89 mm Hg, and PVR of 2.6 WU in the next cardiac catheterization. For these reasons, we decided to occlude

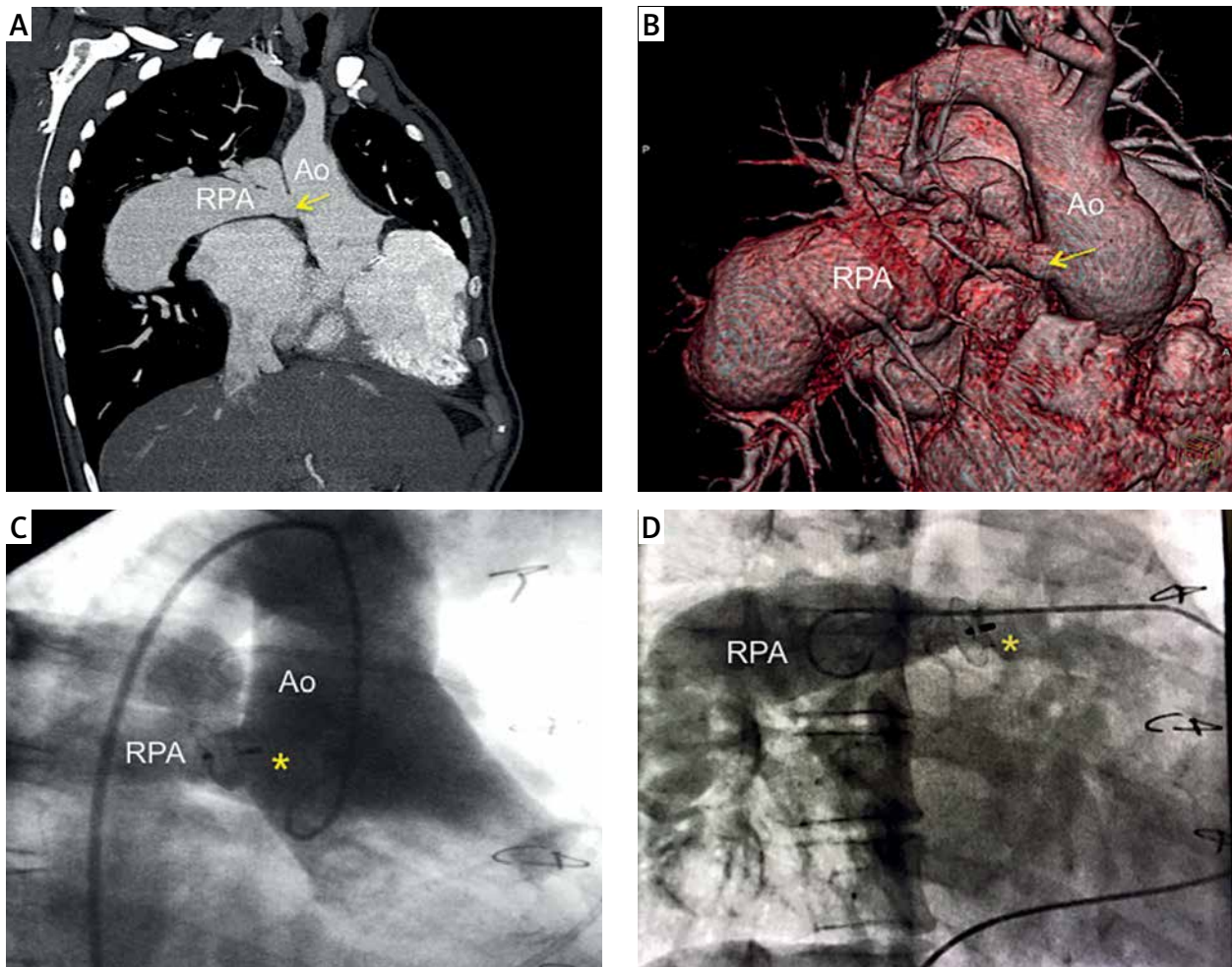
the shunt with a 12 mm Amplatzer muscular ventricular septal defect occluder (MVSO) (Figure 1 C, yellow asterisk) from the arterial side, achieving a reduction of PA trunk and left PA pressure, with a value of 62/28/41 mm Hg. The patient remained asymptomatic until 43 years old, when she was hospitalized due to atrial fibrillation, successfully treated with ablation [3]. The right catheterization showed a PA pressure of 45/20/30 mm Hg, an Ao pressure of 125/84 mm Hg, and a complete closure of the WS (Figure 1 D). The clinical worsening of the patient, and our positive experience with sildenafil for the treatment of elevated pulmonary pressure in a child after the Glenn procedure [4], represented the principal reasons for the shunt closure and subsequent sildenafil therapy. In our opinion, the MVSO with symmetrical retention discs is able to reduce the risk of embolization in case of pulmonary hypertension. Another option of treatment could be implantation of a covered stent in the pulmonary artery to occlude the WS; indeed, an additional advantage of this therapy is the dilation of the stenosed RPA. Our case shows how a combined strategy of medical treatment with sildenafil and percutaneous occlusion of the residual aortic-pulmonary shunt may be useful for the management of pulmonary hypertension in a patient with corrected TOF.

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**Figure 1.** 2D (A) and 3D (B) CT angiogram features of the shunt between the ascending aorta (Ao) and right pulmonary artery (RPA) (yellow arrow). Angiographic results of percutaneous occlusion of aortic-pulmonary shunt with 12 mm Amplatzer MVSO (yellow asterisk) implanted from the arterial side, immediately after the intervention (C) and 5 years later (D)

## Conflict of interest

The authors declare no conflict of interest.

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