Transcatheter therapy of partial anomalous pulmonary venous connection with dual drainage and coarctation of the aorta in a single patient

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A 12-year-old boy was found to have aortic coarctation and a partial anomalous pulmonary venous connection. Historically, multiple cardiac pathologies, such as in the present case, required a surgical approach. We describe transcatheter treatment of the coarctation with a stent and occlusion of the partial anomalous pulmonary venous connection with an Amplatzer vascular plug in a single patient without complications.

Keywords: Coarctation of aorta, Dual drainage, Partial anomalous pulmonary venous connection, Stent, Vascular plug, Vertical vein

1. Introduction

Partial anomalous pulmonary venous connection (PAPVC) is an extremely rare congenital condition where one or more of the pulmonary veins are connected to the venous circulation. Its prevalence within the general population is 0.4–0.7% [1]. Approximately 90% of all PAPVCs originate from the right lung, 7% originate from the left lung, and 3% of patients are found to have bilateral PAPVCs originating from both lungs connecting to the superior vena cava (SVC), the inferior vena cava (IVC), the right atrium, or the innominate vein. Partial anomalous pulmonary venous connections are frequently associated with atrial septal defects [1–3], and are rarely associated with other congenital abnormalities of the heart [4]. Echocardiography is the initial modality of choice for the noninvasive detection of PAPVC [1]. Generally, patients with a partial anomalous pulmonary venous connection, if symptomatic or showing evidence of significant left-to-right shunting, are treated with surgery. Occasionally,

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it is possible to treat partial anomalous pulmonary venous connections percutaneously by means of an Amplatzer occlusion device (AGA Medical Corporation, Plymouth, USA), although this is only feasible when the anomalous pulmonary veins connect both to the left atrium and the systemic veins. To our knowledge, only few such cases have been previously described in the literature [5–7]. We report a case of transcatheter treatment of coarctation of the aorta and a partial anomalous pulmonary venous connection in a single patient.

2. Case report

A 12-year-old boy presented with persistent headache and leg pain for the past 8 months. On examination, he was found to be hypertensive with blood pressure 132/90 and had an ejection systolic murmur. Subsequent echocardiogram revealed evidence of an aortic coarctation beyond the origin of the left subclavian artery and partial anomalous drainage of the left upper lobe pulmonary vein via a vertical vein into the systemic left innominate vein. He underwent cardiac magnetic resonance imaging, which showed coarctation of aorta and left upper pulmonary vein draining to left innominate vein. The patient's magnetic resonance angiogram study of brain showed no cerebral artery aneurysms. The patient underwent cardiac catheterization and angiography revealed left aortic arch with normal branching and a coarctation just at the origin of the left subclavian artery (Fig. 1) with a peak-to-peak gradient of 20 mmHg and a minimum coarctation diameter of 7 mm and poststenotic diameter of 20 mm and the diameter of descending aorta at the level of diaphragm was 16 mm. A 3.4-cm Bare Cheatham Platinum stent (NuMed Inc., Newyork City, USA) mounted on a 20-mm Z-med II balloon (NuMed Inc., Newyork City, USA) was positioned across the coarctation and deployed (Fig. 2), with no residual gradient after stenting.

Innominate vein angiogram showed no left superior vena cava but a significant left-to-right shunt via the anomalous venous connection from the left upper lobe pulmonary vein via a vertical vein into the systemic left innominate vein. The oxygen saturations in the left and right pulmonary vein were 98%, in the left innominate vein 98%, and in the right innominate vein 82%. A repeat procedure was undertaken for occlusion of the partial anomalous pulmonary venous connection. Angiography of the left pulmonary artery showed good size left pulmonary artery and confirmed drainage of the left upper pulmonary vein via a vertical vein into the innominate vein (Fig. 3). Angiography of the right pulmonary artery showed good size right pulmonary artery and all right sided pulmonary veins draining to left atrium and no atrial septal defect. The use of 6F Berman wedge catheter balloon occlusion venogram of the vertical vein showed dual supply of the left upper pulmonary vein, one to a vertical vein and the other to the left lower pulmonary vein which is draining to the left atrium (Fig. 4). While occluding the vertical vein, the innominate vein pressure remained at 8–10 mmHg. A

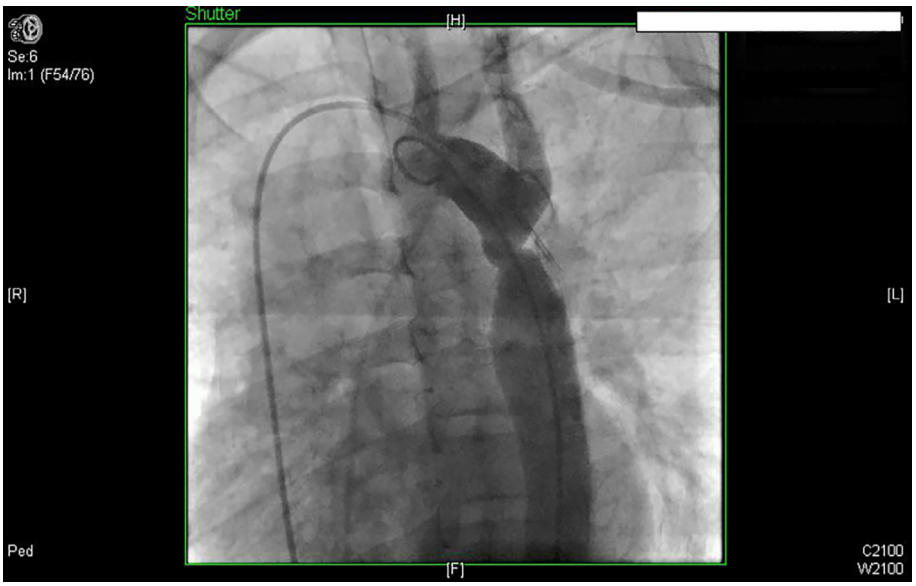

Figure 1. Aortic angiogram illustrating aortic coarctation.

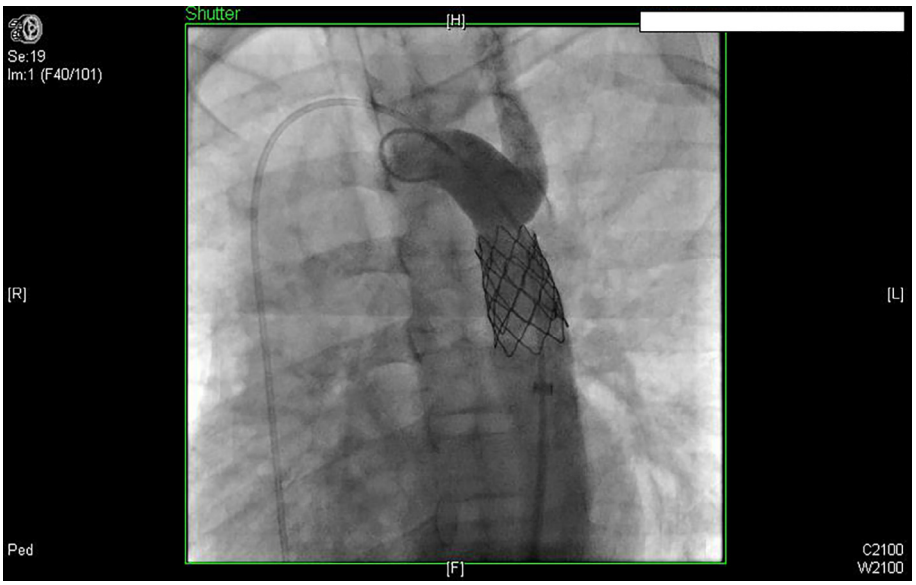

Figure 2. Aortic coarctation after treatment with a 3.4-cm Bare CP stent (NuMed Inc., Newyork City, USA). CP = Cheatham platinum.

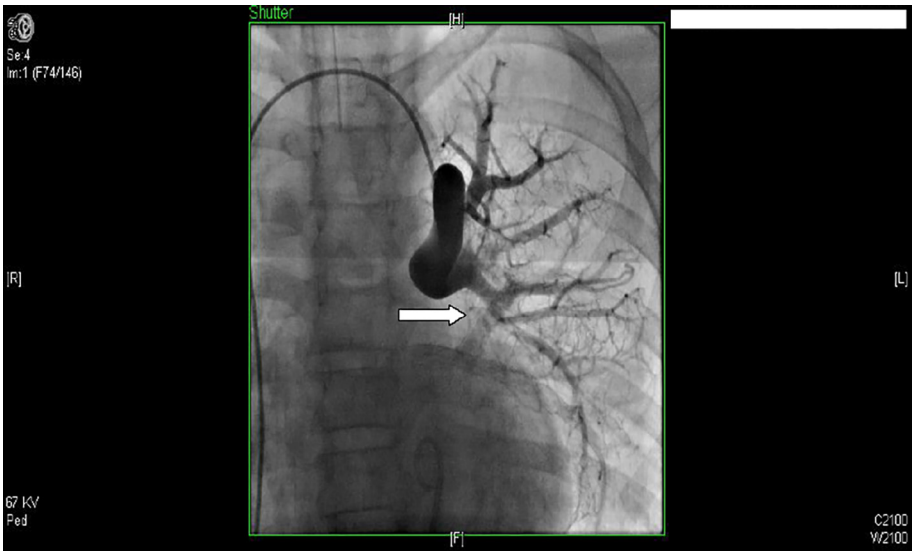

Figure 3. Balloon occlusion venogram of the vertical vein showed dual supply of left upper pulmonary vein showing dual drainage of the left upper pulmonary vein via a vertical vein and the left atrium (arrow).

16 × 12-mm Amplatzer Vascular Plug II (AGA Medical Corporation, Plymouth, USA) was used to occlude the vertical vein before its junction with the innominate vein (Fig. 5). Repeat angiography confirmed no residual leak. The patient remained stable and a chest X-ray revealed that both prostheses remained well seated.

3. Discussion

A partial anomalous pulmonary venous connection occurs when at least one of the pulmonary veins connect to the right atrium directly or indirectly through one or more of its venous tributaries [8]. No definitive classification system has been proposed for this disorder, although Alsoufi et al. [9] have proposed five subtypes of partial anomalous pulmonary venous connection, which include the following: (1) right partial anomalous pulmonary venous connection to the superior caval vein: the most common type, in which the anomalous pulmonary veins attach to the lower side of the superior caval vein or the superior caval vein/right atrium junction; (2) right partial anomalous pulmonary venous connection to the right atrium. The right pulmonary veins connect

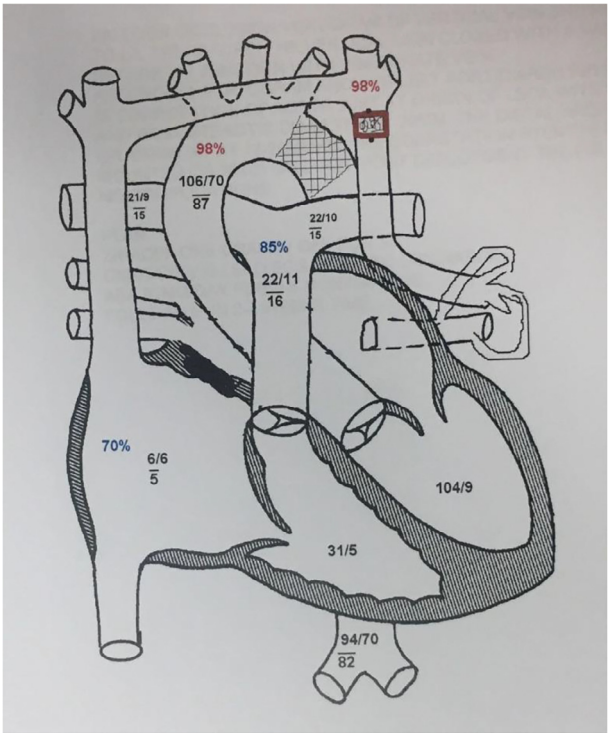

Figure 4. Diagram showing partial anomalous pulmonary venous connection with dual drainage and coarctation of the aorta.

directly to the right atrium; (3) right partial anomalous pulmonary venous connection to the inferior caval vein (scimitar syndrome). The anomalous right pulmonary vein, generally draining the entire right lung, descends in a cephalocaudal direction toward the diaphragm, then joins the inferior caval vein or inferior caval vein/right atrial junction; (4) left partial anomalous pulmonary venous connection. The left pulmonary veins connect to the left innominate vein by way of an anomalous vertical vein; and (5) bilateral partial anomalous pulmonary venous connection; a rare form of partial anomalous pulmonary venous connection. Most commonly, the atrial septum is intact, the left superior pulmonary vein attaches to the left innominate vein by way of an anomalous vertical vein, and the right superior pulmonary vein attaches to the superior caval vein/right atrial junction.

Here we describe a less common, type 4 variety of partial anomalous pulmonary venous connection, in which the anomalous pulmonary vein (vertical vein) ascends into the left innominate vein, in association with coarctation of the aorta, which was discovered during routine assessment. Until more recently, such cases with multiple cardiac pathologies would have been treated using a surgical approach. However, advancement and refinement of percutaneous techniques has meant that such cases involving multiple cardiac pathologies can be treated using interventional percutaneous techniques, obviating the need for surgery with very good outcome data. Endovascular stent implantation has become the modality of choice for the treatment of coarctation of the aorta in older children and adults, and is associated with relatively low morbidity and mortality [10]. The dual drainage allowed successful percutaneous closure of the vertical vein without obstruction to the pulmonary venous flow to the left atrium. To our knowledge, there is only one reported case [7] of coarctation in association with partial anomalous pulmonary venous connection treated successfully with a percutaneous approach, thereby obviating the need for surgery and its associated risks.

In partially abnormal pulmonary venous return with dual drainage, transcatheter therapy can be

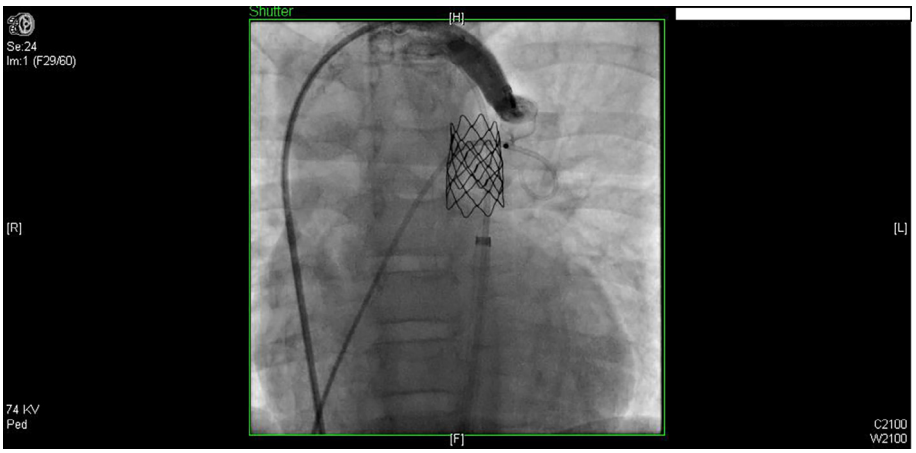

Figure 5. Amplatzer Vascular Plug II (AGA Medical Corporation, Plymouth, USA) was used to occlude the vertical vein before its junction with the innominate vein.

offered in most patients. This case illustrates the feasibility of complete interventional cure of select cases of partial anomalous pulmonary venous drainage with dual drainage. Even in patients with multiple cardiac pathologies, percutaneous treatment options should be considered in close liaison with surgical colleagues before the decision for a surgical approach to treatment is made.

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