



Transcatheter Occlusion of the Vertical Vein in a Partial Anomalous Pulmonary Venous Connection: A Case Report and Literature Review

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Partial anomalous pulmonary venous connection (PAPVC) is a relatively rare congenital heart anomaly characterized by abnormal drainage of one or more pulmonary veins into a location other than the left atrium. In this case report, we present a 70-year-old male diagnosed with PAPVC with a vertical vein connected between the left upper pulmonary vein and the innominate vein. The transcatheter occlusion of the vertical vein was successfully achieved using a vascular plug. The report underscores transcatheter occlusion as a minimally invasive and effective alternative to surgical correction for PAPVC; it may provide advantages such as a shorter hospital stay, faster recovery time, and reduced complications.

Key words: Partial anomalous pulmonary venous connection, vertical vein, transcatheter occlusion, vascular plug

INTRODUCTION

Partial anomalous pulmonary venous connection (PAPVC) is a less frequently occurring congenital anomaly, in which certain pulmonary veins, responsible for transporting oxygenated blood from the lungs to the heart, exhibit abnormal connections to the right atrium or its tributaries instead of the left atrium. The prevalence of partial anomalous pulmonary venous return (PAPVR) is approximately 0.5%.¹ The most frequent anomaly is the anomalous drainage of the right superior pulmonary vein and often associated with sinus venosus atrial septal defect. Left-sided anomaly is less and commonly drains to the left brachiocephalic vein through a vertical vein or to the coronary sinus.²

While the conventional gold standard involves surgically redirecting anomalous pulmonary vein(s) to the left atrium, there is a growing trend and increasing experience in utilizing percutaneous transcatheter device closure as an alternative for specific cases of PAPVR. In this context, we present a successful case that transcatheter closure effectively resolved the anomaly.

CASE REPORT

A 70-year-old male presented with a 1-month history of intermittent chest tightness and palpitations. Upon arrival at the emergency department, his vital signs were stable, and there was no desaturation under ambient oxygen. Electrocardiography revealed sinus rhythm with no ST-segment elevation, and laboratory results indicated normal levels of creatine kinase (54 U/L) and Troponin-I (25 pg/mL). Echocardiography showed normal left ventricular systolic function, an ejection fraction of 61.3%, mild tricuspid regurgitation, and mild dilation of the right ventricle with a systolic pressure of 26 mmHg. Contrast-enhanced computed tomography (CT), conducted for chest pain evaluation, revealed atherosclerosis in the aorta and coronary arteries, along with an incidental finding of an anomalous left upper pulmonary vein draining into the left innominate vein. Right heart catheterization demonstrated a mildly increased pulmonary artery pressure of 33/15 mmHg and elevated oxygen saturation in the superior vena cava

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(84.9%). Angiography confirmed the anomalous drainage, depicting the left upper pulmonary vein connecting to the left innominate vein through a vertical vessel [Figure 1]. Coronary catheterization identified tubular stenosis (approximately 70%) in the middle of the posterior descending artery and discrete stenosis in the middle third of the left circumflex and proximal obtuse marginal coronary artery. Attempts at transluminal coronary angioplasty with trek balloons revealed a residual 50% stenosis and a type B dissection, based on the National Heart, Lung, and Blood Institute system.³ Subsequently, a 2.5 mm × 23 mm combo stent was deployed, resulting in no residual stenosis upon final angiography. The patient received dual antiplatelet medications postprocedure.

One month later, he underwent intervention for the partial anomalous pulmonary vein connection. Cardiac catheterization was performed under local anesthesia using a 6 Fr sheath through right femoral vein access. A 0.035 wire was advanced to the left anomalous vertical vein, connected to the left upper pulmonary vein, and confirmed by angiography. Subsequently, a 90 cm shuttle sheath was exchanged. The minimal diameter of the vertical vein was measured at approximately 1.1 cm. Therefore, we selected a 16 mm (Vascular Plug Diameter) and 12 mm (Unconstrained Length) Amplatzer Vascular Plug II (Abbott Vascular, IL, USA) to occlude the vertical vein. The final position, confirmed by angiography [Figure 2], did not interfere with the flow of the innominate vein. The hospital course was uneventful, and the patient was discharged the next day. At the 1-year follow-up, the patient reported no occasional dizziness, chest tightness, or any embolic events. A repeat contrast-enhanced CT obtained 8 months later showed satisfactory results [Figure 3].

DISCUSSION

The anomalous connection or drainage of pulmonary



Figure 1: Angiography revealed the left upper pulmonary vein draining into the left innominate vein. Through a vertical vessel

veins arises from the failure of one or more pulmonary veins to establish a connection with the left atrium during fetal development. Different levels of dilation in the right ventricle and right atrium are typically observed due to a persistent left-to-right shunt, leading to increased blood flow to the lungs and potential remodeling of the pulmonary vascular bed, which can result in pulmonary arterial hypertension. In cases of this pathology, the conventional approach involves surgically redirecting the anomalous vein(s) to the left atrium, considered the gold standard procedure.⁴

While transthoracic echocardiography can offer valuable insights into cardiac structure and function, its capacity to outline the intricate anatomy of anomalous veins in PAPVC may be constrained. Advanced imaging modalities such as cardiac magnetic resonance imaging and CT angiography excel in providing detailed information about extracardiac vascular anatomy. In addition, cardiac catheterization proves useful in assessing defined hemodynamics.⁵

Surgical correction may be recommended to prevent potential complications and to alleviate the impact of the left-to-right shunt on cardiac function.⁵ In asymptomatic patients, the indication for intervention lies in the presence of evidence indicating volume overload, such as right heart dilation, progressive tricuspid regurgitation, or early stages of pulmonary vascular disease. The choice between surgical repair, which may involve median sternotomy or thoracotomy, with or without cardiopulmonary bypass support, depends on the specifics of individual cases.⁶

Complications associated with surgical repair encompass issues such as kinking, stenosis, and thrombosis. Pulmonary venous obstruction, characterized by a substantial gradient, may result in right-heart enlargement, recurrent left-sided pneumonia, thrombosis, arrhythmias, and pleural effusions. A specific literature source indicates that while surgical

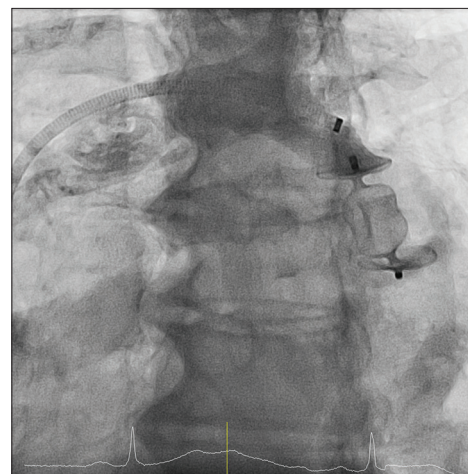


Figure 2: After the deployment of the Amplatzer Vascular Plug, angiography indicated a satisfactory. Positioning of the vascular plug, without any interference with the innominate vein

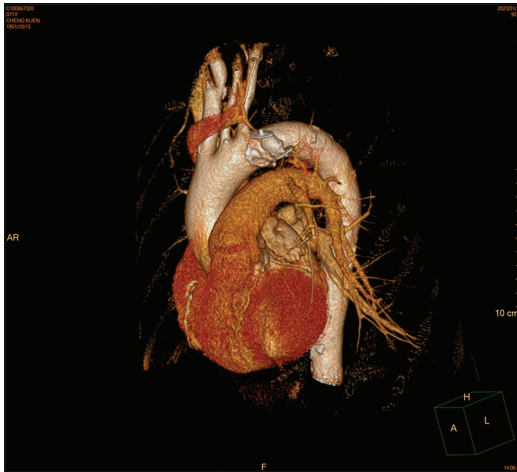


Figure 3: A follow-up contrast-enhanced computed tomography, conducted 8 months later, demonstrated a favorable position without any impingement on any structure and total occlusion of the vertical vein

treatment is associated with low time-related morbidity, there is an elevated risk of postoperative pulmonary venous obstruction and abnormally diminished perfusion of the right lung in the management of children with scimitar syndrome.⁴

PAPVR may exhibit single drainage (to the systemic veins) or dual drainage (to both the systemic veins and the left atrium). The choice of management is contingent upon the impact of the shunt on the heart and lungs, with surgical correction being the typical approach. In instances of PAPVR with dual drainage as in this presented case, there is a contemporary approach involving percutaneous occlusion of the vertical vein with dual drainage anatomy. This technique redirects blood flow, compelling it to flow into the left atrium, mimicking the normal cardiac circulation pattern.

In contemporary times, the advancement of endovascular techniques allows for a less invasive approach to correct the anomaly. Some pediatric cases undergoing transcatheter occlusion for PAPVR have demonstrated fast recovery, shorter in-hospital stays, lower costs, and heightened patient satisfaction.⁷

CONCLUSION

In this case report, the transcatheter occlusion of the vertical vein in PAPVR has proven to be a viable alternative to surgical treatment. It is associated with effectiveness, safety, shorter hospital stays, and faster recovery times. However, to establish its wide utilization, extensive studies on a large scale and long-term follow-ups are imperative.

Ethical approval statement

The study was conducted in accordance with the Declaration of Helsinki and was approved by the Institutional Review Board of Tri-Service General Hospital (approval number: B202415128; approval date: 2024/06/03).

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In this form, the patient gave his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Data availability statement

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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