

# Trans-catheter closure of ASD and abnormal connection of left pulmonary vein to vertical vein

## A case report

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### Abstract

**Rationale:** Partial anomalous pulmonary venous connection is a rare congenital anomaly in which one or more pulmonary veins are connected to the venous circulation leading to left to right heart shunt. Although correction of anomalous pulmonary venous connection is achieved through surgery, there are rare instances where the abnormal pulmonary vein has dual connection to both left atrium and the major systemic veins. Under these circumstances, catheter-based treatment might become a feasible option.

**Patient concerns:** A 22-year-old female presented with exertional dyspnea, holo-systolic murmur in left sternal border, and fixed splitting of S2 in examination.

**Diagnosis:** The patient was diagnosed with secundum type atrial septal defect (ASD) and dual drainage of left upper pulmonary vein.

**Interventions:** The patient was candidate for device closure. Under TEE guidance, occluder devices were deployed in the upper part of vertical vein and subsequently in place of ASD.

**Outcomes:** Echocardiogram in the next day showed complete occlusion of flow through the vertical vein and ASD. Dual antiplatelet was prescribed on discharge. Follow-up echocardiography after 3 months showed obvious improvement in RV size. Due to suspicion for clot formation, TEE was done and thrombosis with approximate length of extension of 15 mm was detected back to the device. The patient is following for 5 years. Repeated TEE after 2 years did not show any change in the burden of clot.

**Lessons:** For comprehensive evaluation of patients with ASD, assessment of pulmonic veins is crucial and in the presence of a vertical vein, the dual drainage of pulmonic veins should be considered.

**Abbreviations:** ASD = atrial septal defect, LUPV = left upper pulmonary veins, PAPVC = partial anomalous pulmonary venous connection, TEE = transesophageal echocardiography, TTE = transthoracic echocardiography, VSD = ventricular septal defect.

**Keywords:** device closure, partial anomalous pulmonary venous connection, vertical vein

## 1. Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital anomaly in which pulmonary veins carry blood from the lungs to the right side of the heart. The condition has a prevalence of 0.4% to 0.7%, it is frequently diagnosed as an incidental finding.<sup>[1,2]</sup> Herein we described an adult patient with atrial septal defect (ASD), in which the left upper pulmonary veins (LUPV) drained into the innominate vein which were successfully obstructed by occlusion device.

## 2. Case presentation

A 22-year-old female was referred to our clinic for exertional dyspnea since one month ago. Physical examination defined holo-systolic murmur in left lower sternal border with fixed splitting of S2. Twelve-lead electrocardiogram showed normal sinus rhythm with incomplete right bundle branch block. Transthoracic echocardiography (TTE) was done. Right ventricle was dilated with preserved systolic function. Paradoxical septal motion was noted. There was mild to moderate tricuspid

Written consent to publish this information was obtained from study participant.

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

The data that support the findings of this study are available from Imam Reza Hospital, Mashhad University of Medical Sciences, but restrictions apply to the availability of these data, which were used under license for the current study, and so are not publicly available. Data are however available from the authors upon reasonable request and with permission of the corresponding author.

Publishing this case presentation is performed according to ethical guidelines of Mashhad University of Medical Sciences. Patient was evaluated for his problem and fulfilled informed consent for participation.

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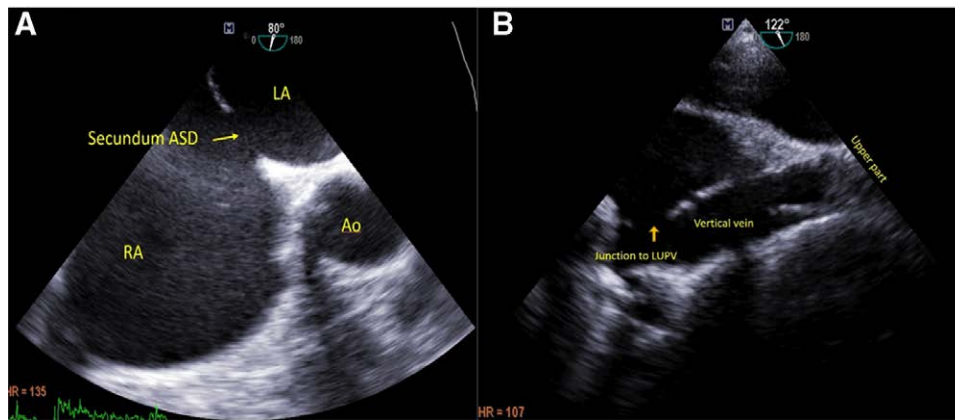
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**Figure 1.** Transesophageal echocardiography images. (a) Two-dimensional TEE image shows atrial septal defect and (b) vertical vein before device closure. Ao = aorta, LA = left atrium, LUPV = left upper pulmonary vein, RA = right atrium, TEE = trans esophageal echocardiography.

regurgitation. Secundum ASD was seen with left to right shunt. The estimated systolic pulmonary artery pressure was 45 mmHg. A vertical vein was detected traversing at the left side of the thoracic descending aorta and draining into innominate vein. The calculated Qp/Qs was 1.85. In transesophageal echocardiography (TEE), ASD size was 16 mm, with suitable rims (except to the antero-superior rim). Left and right pulmonary veins were seen draining into left atrium. LUPV seemed to be narrow with turbulent flow passing the vein (Peak velocity: 1.2 m/s, mean pressure gradient: 5 mmHg). The vertical vein and the abnormal drainage of LUPV could be clearly seen in TEE (Fig. 1). Diameter of LUPV and the site of connection to the vertical vein was about 5-mm in 2D TEE. No other associated abnormality was reported. The patient underwent cardiac catheterization that confirmed secundum type ASD and the connection of the vertical vein to both innominate vein and the left atrium via the left upper pulmonary vein (Fig. 2).

The patient was referred for device closure. On admission blood pressure was 110/60 mmHg, heart rate was 85 beats/min, and respiratory rate was 18 breaths/min and O<sub>2</sub> saturation at room temperature was 94%. After local anesthesia and placing two 6F sheaths in femoral artery and femoral vein, full oximetry run was done. Left to right shunts in level of pulmonary vein to abnormal connection and in ASD level was proved. The patient deeply sedated. Then 6f sheath of vein access was replaced with 10F sheath. Under TEE guidance, muscular ventricular septal defect occluder (12mm) was deployed in the upper part of vertical vein, at its connection to innominate vein (Fig. 3a). Contrast injection proved optimal occlusion. Then, under TEE guidance and using the same 10F sheath, ASD occluder (Figulla® Flex II ASD, 18 mm) was deployed in place of ASD (Fig. 3b). TEE proved eliminated flow of the vertical vein and proper position of devices with no compressive effect on adjacent structures and no clot (Fig. 4).

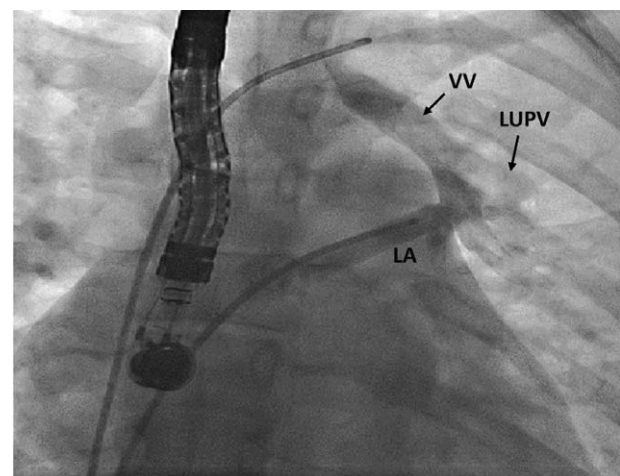
Follow-up echocardiogram the next day showed complete occlusion of flow through the vertical vein and ASD. Dual antiplatelet with ASA and Clopidogrel were prescribed on discharge.

Trans-thoracic echocardiography was done three months later. There was obvious improvement in right ventricle size. Haziness was suspected behind the device in the vertical vein, which was confirmed in TEE, and ascribed to the clot formation back to the device with approximate length of 15 mm (Fig. 5). Turbulent flow in the narrow LUPV was seen with peak velocity of flow reaching to 1.7 m/s. Pulmonary artery pressure was normal (#20 mmHg). The patient is following for 5 years. She is symptom free. Repeated TEE after 2 years did not show any change in the burden of clot. Single antiplatelet therapy with ASA continued. On annual TTE, the turbulent flow in LUPV was still evident. Pulmonary artery pressure remained in normal limits.

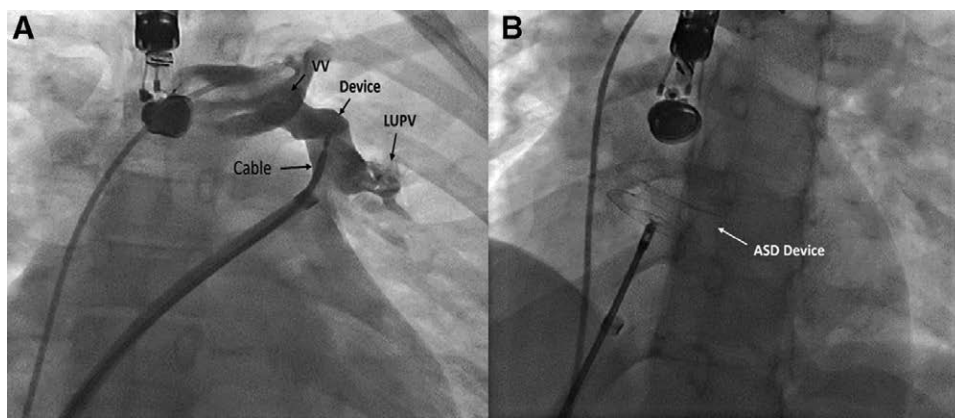
### 3. Discussion

PAPVC is a relatively uncommon congenital anomaly characterized by one or more pulmonary veins draining into a systemic vein or the right atrium rather than the left atrium. Overall, PAPVC is symptomatic and often co-exists with ASD, in 80% to 90% of the cases.<sup>[3]</sup> Dual drainage of the pulmonary veins to both a systemic vein and LA is rare and difficult to estimating the incidence, because most of these patients are asymptomatic.<sup>[4]</sup> Trans-catheter closure of the abnormal venous connection is certainly the treatment of choice for PAPVC with dual drainage, although successful surgical ligation of the vertical vein has been reported.<sup>[5]</sup> The closure could be attained by coil or Amplatzer duct occluder or vascular plugs.<sup>[6]</sup> The type of closure device used is determined by the anatomical size and shape of the abnormal venous connection.<sup>[4]</sup> We used a muscular ventricular septal defect occluder in our patient providing complete occlusion of vertical vein; concurrently utilized Amplatzer® occluder for secundum ASD. Performing a balloon occlusion test in the vertical vein, before implanting the device has been suggested to confirm the adequacy of the alternative drainage. Device closure could not be done in cases with  $\geq 10$  mmHg increase in pulmonary venous pressure.

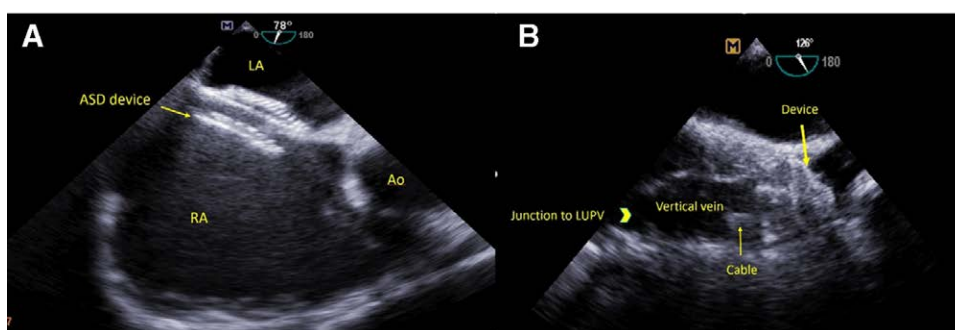
In this case, the flow velocity of pulmonic vein was increased from 1.2 to 1.7 m/s after the device closure; but pulmonary



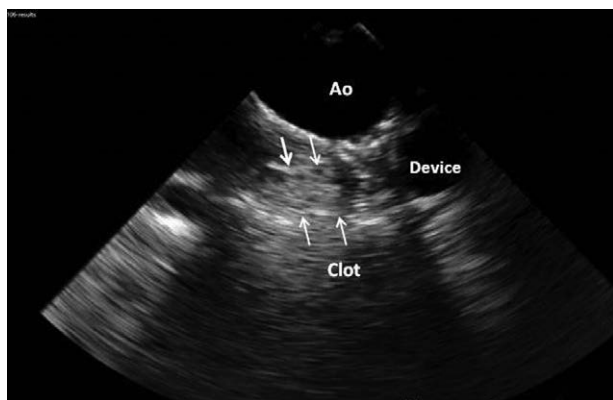
**Figure 2.** The catheter was positioned in the left pulmonary vein, where the angiography was performed. LA = left atrium, LUPV = left upper pulmonary vein, VV = vertical vein.



**Figure 3.** Angiogram demonstrate (a) VSD occluder device in proper position in the abnormal connection without significant residual flow into the left atrium and (b) ASD device. ASD = atrial septal defect, LUPV = left upper pulmonary vein, VSD = ventricular septal defect, VV = vertical vein.



**Figure 4.** Transesophageal echocardiography images. The images show (a) the ASD device deployed in an appropriate position and (b) the VSD occluder device with complete occlusion of the vertical vein. Ao: aorta, ASD = atrial septal defect, LA = left atrium, LUPV = left upper pulmonary vein, RA = right atrium, VSD = ventricular septal defect.



**Figure 5.** Transesophageal echocardiography images. It shows the clot formation back to the device in the vertical vein.

artery pressure did not increase during 5-year follow up. Device thrombosis would be a possible serious complication. Optimal antiplatelet or antithrombotic medication have been proposed to enhance safe and complete endothelial coverage of the implanted device. We encountered clot formation behind the implanted device while the patient was on dual antiplatelet therapy, which was ascribed to stagnation of flow. Conservative management with periodic follow up was selected and there was not further extension or increase in the burden of clot on periodic TTE studies. The patient is still receiving ASA a single antiplatelet therapy.<sup>[7]</sup>

Although TTE is usually unable to detect thrombus formation on the device, routine TEE examination is not suggested in adult

patients in many centers, and TTE as an imaging tool might be sufficient. TEE could be performed in cases when transthoracic echocardiography suggests thrombosis or when transthoracic images are suboptimal.<sup>[7]</sup>

#### 4. Conclusion

For comprehensive evaluation of patients with ASD, assessment of pulmonic veins is crucial and in the presence of a vertical vein, the dual drainage of pulmonic veins should be considered. Trans-catheter interventions could be a safe and efficient treatment of such cases and the thrombus formation after device closure of the vertical vein could be managed conservatively.

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#### Author contributions

M.M.S. and H.P. analyzed and interpreted the patient data regarding the cardiovascular disease and managed the patient. H.P. wrote the first draft of the manuscript. A.K. helped in data gathering and diagnosis patient in echocardiographic evaluation. A.E. helped in patient catheterization and management. F.K. analyzed and interpreted the patient data and wrote the first draft of the manuscript. All authors read and approved the final manuscript.

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