

# A rare case on anomalous pulmonary venous drainage in congenital cardiovascular disease

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## Abstract

anomalous pulmonary veins drain into the right side of the left atrium is an uncommon variety of anomalous pulmonary venous return. Rarely, anomalous pulmonary venous drainage combined with cor triatriatum and atrial septal defect. We presented the imaging findings of a male patient who had anomalous pulmonary venous drainage which has not previously been described.

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**KEYWORDS:** atrial septal defect, cor triatriatum, anomalous pulmonary venous drainage

## 1 | INTRODUCTION

This is the first report of rare simultaneous complication of three cardiac malformations: atrial septal defect, cor triatriatum and anomalous pulmonary veins drain into the right side of the left atrium. A single dilated and tortuous pulmonary vein at the right side draining into the left atrium is extremely rare and has not previously been described. We successfully operated to repair the atrial septal defect and correct the cor triatriatum. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the Scientific Research Ethics Committee of Qilu Hospital of Shandong University and individual consent was waived in accordance with local policies about case reports.

### | Case report

An 18-year-old male patient presented with a systolic murmur in the second and third intercostal spaces at the left sternal border during physical examinations. His transthoracic echocardiogram showed an atrial septal defect (ASD) with left to right shunt and unclear drainage of pulmonary veins. Moreover, there was

a deformed septum in the left atrium, which divided the left atrium into two parts (the true left atrium and an accessory atrium) communicating through a 7mm-diameter hole. The echocardiogram showed that preoperative pulmonary artery systolic pressure was about 49 mmHg.

We successfully operated on the patient for closure of the ASD and correction of the cor triatriatum by removing the deformed septum in the left atrium. However, the patient was explored with a single tortuous pulmonary vein draining into the top right corner of the left atrium during surgery. Nine months after surgery, the patient underwent computerized tomography angiography (CTA) for follow-up study. Review of CTA images revealed the presence of a single dilated and tortuous pulmonary vein at the right side draining into the left atrium. In addition, a second pulmonary vein at the left side was found to directly merge into the right-sided pulmonary vein instead of connection with the left atrium (Figure 1). The transthoracic echocardiogram showed that pulmonary artery systolic pressure was about 25 mmHg after surgery.

## 2 | DISCUSSION

Both sides of pulmonary veins with a single opening in the left atrium presented in this case is a rare variety of anomalous pulmonary venous drainage, which is very common in patients with heterotaxia. But in patients with normal visceratrial situs, it usually occurs as an isolated deformity and barely combined with other complicated congenital heart diseases such as cor triatriatum<sup>1</sup>. Previous studies showed that anomalous drainage of left-sided pulmonary veins is more common than right-sided pulmonary veins<sup>2</sup>. To the best of our knowledge, this is the first report of anomalous pulmonary venous drainage combined with cor triatriatum and atrial septal defect. The goal of corrective surgery in patients with anomalous pulmonary venous drainage, is to eliminate left to right shunt, pulmonary hypertension and re-establish the structure of the atrium<sup>3</sup>. With the pattern of anomalous pulmonary venous drainage shown in our patient, we decided not to correct the dilated and tortuous right-sided pulmonary vein because the deformed vein can normally drain into left atrium and the surgery also can reduce the pulmonary artery systolic pressure that bring us good clinical outcomes. The success of this procedure might provide a valuable reference for surgeons in the future clinical practice. For those patients with anomalous pulmonary venous drainage into the left atrium as well as cor triatriatum and atrial septal defects, the pulmonary artery pressure could be decreased only by repairing the atrial defect and removing the diaphragm in the left atrium if the patients have moderate pulmonary hypertension (PAH) before surgery. Furthermore, pulmonary vein CTA can provide more precise anatomical details to help confirm the clinical diagnosis.

## 3| CONCLUSION

Both sides of pulmonary veins with a single opening in the left atrium combined with other complex congenital heart diseases is rarely. The variety of anomalous pulmonary venous drainage is an extremely rare abnormality. CTA is very useful to accurately determine the paths of drainage of pulmonary veins<sup>3</sup>.

## AUTHOR CONTRIBUTIONS

Changcun Fang performed the surgery. Zhuangzhuang Lu and Xiao Bai prepared the manuscript. Guangmin Song and Changcun Fang revised the manuscript critically. All authors read and approved the final manuscript.

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## CONFLICT OF INTERESTS

The authors report no conflicts.

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