

Anomalous Connection of the Left Pulmonary Vein to the Coronary Sinus with Intact Atrial Septum in a Young Woman

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ABSTRACT

Background: Left-sided partial anomalous pulmonary venous connection (PAPVC) is a rare congenital abnormal cardiac defect. An intact atrial septum is more uncommon. As we know, a connection of the left pulmonary vein (LPV) to the coronary sinus (CS) with an intact atrial septum has not been previously reported.

Case report: We report an 18-year-old woman with this rare anomaly. She showed no obvious clinical symptoms. An echocardiogram revealed the primary diagnosis, and this diagnosis was confirmed during operation. This patient underwent a successful surgical repair. Artificial atrial septal defect (ASD) and coronary sinus orifice were inserted into the left atrium by patch. The patient recovered smoothly without complications after the operation.

Conclusion: Given the high risk of developing congestive heart failure, we advocate for intervention at the preschool age. Surgical techniques depend on the number and location of abnormal veins or veins.

INTRODUCTION

Partial anomalous pulmonary venous connection (PAPVC) is a relatively rare congenital heart disease in which partial pulmonary vein blood flows directly or indirectly into the right atrium [Elami 1995]. Drainage of partial pulmonary veins to the right atrium can lead to pulmonary hypertension, an enlarged right atrium, tricuspid regurgitation, and right ventricular dysfunction. The most common presentation (90%) is accompanied with atrial septal defect (ASD) [Broy 2008]. When accompanied with atrial septal defect (ASD), the patient may have obvious symptoms and appear in early childhood. However, PAPVC with an intact atrial septum, the patient may have no obvious clinical symptoms. PAPVC with

an intact atrial septum is rarely reported [Zhang 2016]. As we all know, anomalous connection of the left pulmonary vein to the coronary sinus with intact atrial septum has never been reported.

CASE REPORT

An 18-year-old woman was admitted to the local hospital because of heart murmur found by physical examination. She received an echocardiographic examination that revealed a congenital heart disease. She had no cyanosis and was clinically asymptomatic, however, a grade two systolic murmur on the second and third left intercostal space was audible. She came to our hospital for further evaluation. Electrocardiogram showed mild enlargement of the right atrium and right ventricle, and the diameter of the main pulmonary artery was enlarged, but there was no ASD or tricuspid regurgitation to create the right ventricular system enlargement. (Figure 1) Further ultrasonic examination found an enlarged coronary sinus measuring 2.31 cm in diameter with two left pulmonary veins connecting and meanwhile, the position of right pulmonary vein was normal. (Figure 2) PAPVC associated without ASD of the left pulmonary vein to the coronary sinus was diagnosed by the cardiac ultrasound, and cardiac surgery was confirmed.

The patient underwent successful heart surgery, and the operation was performed under cardiopulmonary bypass. During the operation, the opening of the coronary vein was widened, with the probe reaching the left superior and inferior pulmonary veins via the orifice of the coronary vein sinus. There was an intact atrial septum, and the right ventricle and right atrium were mildly enlarged. The connection of the right pulmonary veins was normal. An artificial atrial septal defect was formed by cutting the atrial septum. Artificial atrial septal defect and coronary sinus containing the left pulmonary vein orifice were patched into left atrial side. The patient recovered smoothly and was discharged eight days later.

Our hospital's ethics committee approved publication of this case report, and written patient consent was obtained.

DISCUSSION

PAPVC is a rare congenital heart disease first reported by Winslow in 1789, and pathological studies show disease

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incidence in 0.7% of the population [Mei 2017]. Research on the incidence of PAPVC has yielded a mixed bag of results because many patients without clinical symptoms were not captured by the PAPVC rate [Kim 2012]. Thus, the real incidence of PAPVC is slightly higher than expected. Most cases of PAPVC are from the right lung, and only 10%–18% of cases of PAPVC from the left pulmonary are reported. PAPVC originating from the right pulmonary occurs 10 times more commonly than the connecting veins starting from the left lung [Mei 2017]. Few studies of anomalous connection of the left pulmonary vein to the coronary venous sinus have been reported [Ho 2014]. And meanwhile, PAPVC presented with ASD was reported in 80%-90% of cases [Elami 1995]. Isolated PAPVC associated without ASD rarely have been reported. Additionally, to our knowledge, left pulmonary vein

draining into the coronary venous sinus with an intact atrial septum has not been reported. The patient was confirmed by echocardiography, and surgical treatment was performed in time. The operation on her successfully was done. The key to a successful operation is that the artificial atrial septal defect and the mesh should be large enough to prevent the occurrence of blood flow obstruction.

The appearance of the clinical symptoms usually occurs in PAPVC during adulthood [Broy 2008]. The pathophysiology of PAPVC is similar to the left-to-right shunt in simple ASD. Image examinations to diagnose PAPVC consist of chest pain films, CT, MRI, and cardiac catheterization [Clarke 2017]. Doppler echocardiography reveals the trend and distribution of pulmonary veins. Therefore, cardiac ultrasonography is becoming the simple and effective method of the PAPVC.

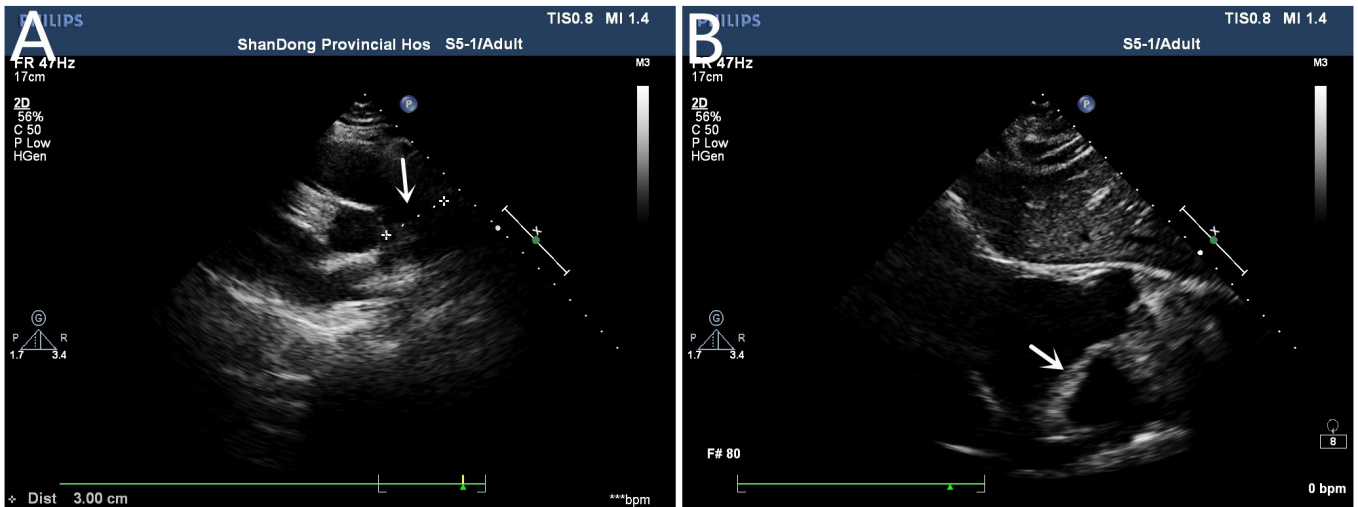


Figure 1. The image analysis by echocardiography. (A) The diameter of the main pulmonary is enlarged (arrows). (B) Demonstrating an intact atrial septum between the right and left atria (arrows).

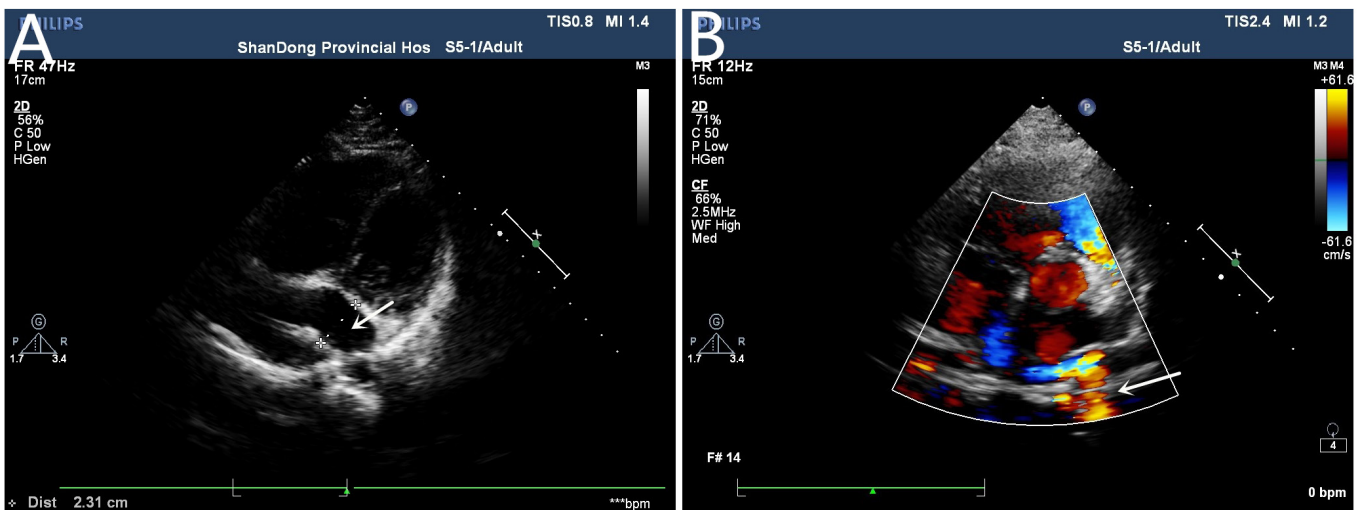


Figure 2. Further analysis by echocardiography. (A) Showing the coronary sinus is widened (arrows). (B) Illustrating anomalous connection of the left pulmonary vein to the coronary sinus (arrows).

Those with PAPVC without symptoms should have a physical exam and an echocardiogram once a year. Patients with the symptoms of heart failure can be controlled with after-load reduction, beta-blockers, diuretics, and cardiac glycosides. Surgical repair was the most effective treatment for PAPVC [Ho 2014]. Preschool age is the optimal time for an intervention. Surgical techniques depend on the number and location of abnormal veins.

CONCLUSION

In this case, by using echocardiography, we were able to detect early, asymptomatic stages of the rare congenital heart condition. The patient successfully was treated with surgery. In view of the above and many other similar reports, we recommend timely surgical treatment to correct the deformity, which is the least risky and has the best results.

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