

CASE REPORT

Surgical treatment of partial anomalous pulmonary venous connection misdiagnosed as atrial septal defect underwent transcatheter occlusion: A case report

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Abstract

This case highlights the importance of coordinating with cardiopediatricians or congenitalists in the evaluation and treatment of ASD.

KEYWORDS

closure, complications, congenital heart disease, surgery

1 | INTRODUCTION

We here report a patient with partial anomalous pulmonary venous connection who was misdiagnosed as ASD and subsequently underwent transcatheter occlusion in another hospital. Two years later, he came to our hospital due to chest tightness and residual shunt. An Amplatzer occluder was removed, and PAPVC correction was performed accordingly.

Partial anomalous pulmonary venous connection (PAPVC) is a rare clinical situation with approximately 0.4%–0.7% in the general population.¹ In patients with PAPVC, echocardiography may not suffice establishing the diagnosis of anomalous pulmonary veins, especially in adult. The patient may be misdiagnosed and refer for wrong treatment when comprehensive examination is not fully performed. We

report a patient with PAPVC who was misdiagnosed as atrial septal defect (ASD) and underwent transcatheter occlusion two years ago in another hospital. The patient was admitted to our department 2 years later with chest tightness and residual shunt. The Amplatzer occluder was removed, and PAPVC was corrected.

2 | CASE REPORT

A 19-year-old male patient who presented with progressive chest tightness and shortness of breath was admitted to our department. He was diagnosed as ASD and treated with transcatheter occlusion two years ago in another hospital. Vital signs were blood pressure: 123/77 mm Hg and heart rate:

The institution at which the work was performed: State Key Laboratory of Cardiovascular Disease, Fuwai Hospital, National Center for Cardiovascular Diseases, Chinese Academy of Medical Sciences, and Peking Union Medical College.

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70 bpm. Physical examination revealed a systolic murmur (2/6) along upper left border of sternum. The jugular venous pressure was normal. Holter showed sinus arrhythmia with occasional atrial extrasystole. Transthoracic echocardiography (TTE) showed that the left ventricular end-diastolic diameter (LVEDD) was 41 mm, right ventricular end-diastolic anteroposterior diameter (RVED) was 29 mm, left ventricular ejection fraction (LVEF) was 65%, and the lower edge of the Amplatzer occluder was detached from the atrial septum. The breach was about 9mm (Video 1). Due to the vague information revealed by TTE, the patient referred CT scan for further examination, which suggested PAPVC and a small secundum ASD (Figure 1).

After diagnosis, the Amplatzer occluder was removed and PAPVC correction was performed under general anesthesia. After general anesthesia, vital signs were blood pressure: 103/62 mm Hg; heart rate: 65 bpm; central venous pressure: 5 mmH₂O; and hemoglobin: 168 g/L. During the operation, we found there was about 10mm leakage at the lower edge

of the Amplatzer occluder. The right inferior and the right superior pulmonary veins were diverted into the right atrium and combined with a 30 × 20 mm secundum ASD (Figure 2). After removing the Amplatzer occluder (Figure 3), a Dacron patch was used to repair the ASD and the right inferior and superior pulmonary veins were separated into the left atrium (Figure 4). After operation, the patient recovered

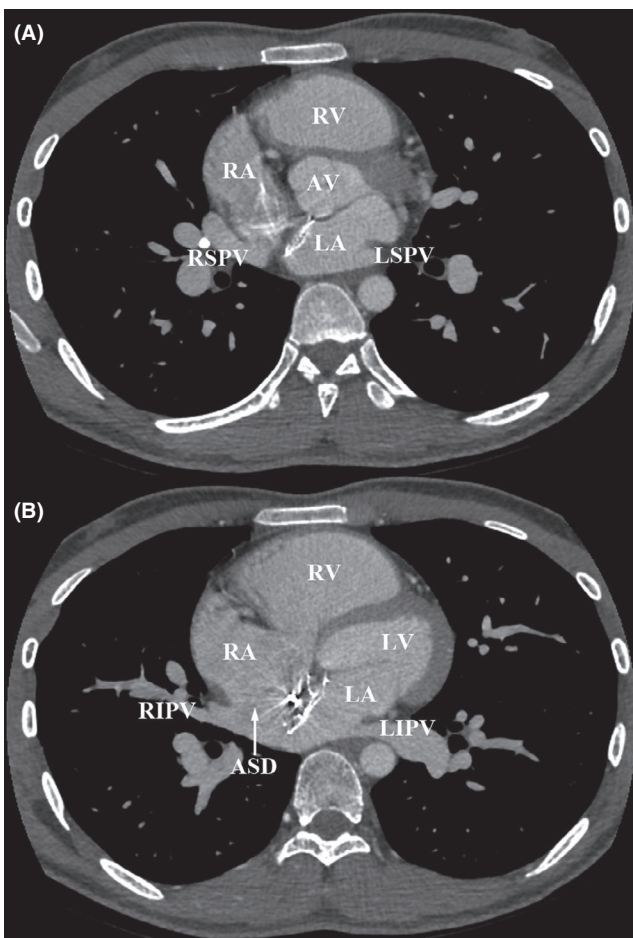


FIGURE 1 Abbreviations: ASD, atrial septal defect; AV, aortic valve; LA, left atrium; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; LV, left ventricular; RA, right atrium; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein; RV: right ventricular

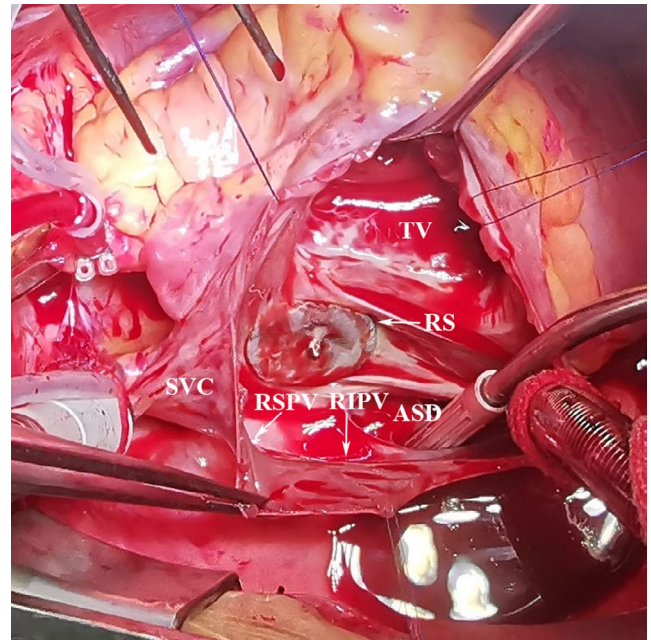


FIGURE 2 Abbreviations: ASD, atrial septal defect; RIPV, right inferior pulmonary vein; RS, residual shunt; RSPV, right superior pulmonary vein; SVC, superior vena cava; TV, tricuspid valve

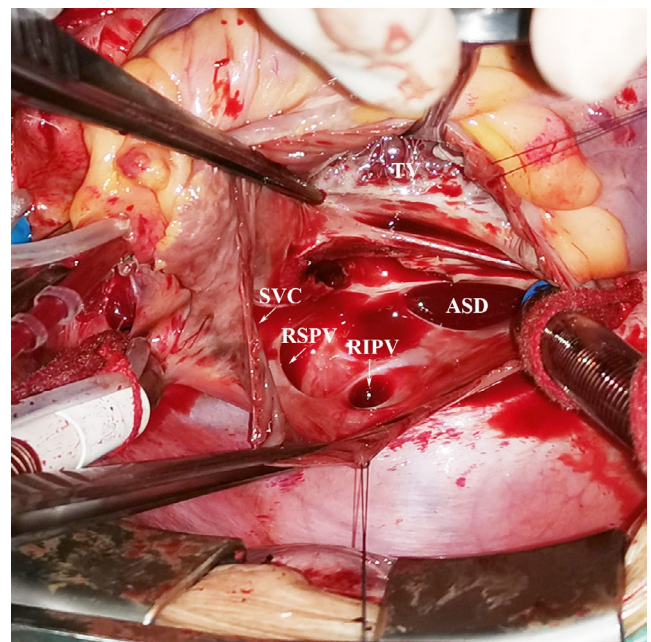


FIGURE 3 Abbreviations: ASD, atrial septal defect; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein; SVC, superior vena cava; TV, tricuspid valve

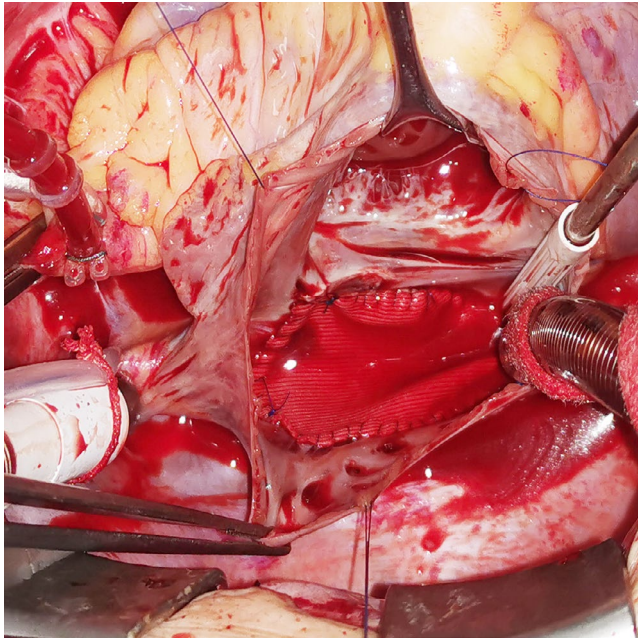


FIGURE 4 After the repair of atrial septum

well. Echocardiography showed that the LVEDD was 48mm, RVED was 20 mm, LVEF was 63%, the pulmonary venous connection was normal, and interatrial shunting was disappeared. After a follow-up of 1 year, the patient has been found to be doing well. TTE showed that the LVEDD was 48 mm, RVED was 24mm, LVEF was 73%, the pulmonary venous connection was normal, and interatrial shunting was disappeared.

3 | DISCUSSION

PAPVC was first reported by Winslow in 1739.¹ PAPVC is a congenital malformation in which 1-3 of the 4 pulmonary veins are connected to the right atrium or systemic vein. Most PAPVC occurs in the right pulmonary vein, as described in this case. PAPVC may occur in isolation or in combination with other congenital heart diseases. PAPVC occurs in approximately 10%-15% of patients with secundum ASDs and in 85% of patients with sinus venosus ASD.²

Significant physiological changes will occur in patients with PAPVC. Irreversible changes of pulmonary vessels will occur if not corrected in time. Therefore, patients with PAPVC had clear indication for surgery. Occasionally, the existence of a dual drainage connecting the PAPVC to the left atrium has been reported.³⁻⁹ In such patients, transcatheter therapy may be an alternative to surgery. However, surgical repair is recommended for patients with PAPVC when functional capacity is impaired and RV enlargement is present, there is a net left-to-right shunt sufficiently large to cause physiological sequelae, PA systolic pressure is less than 50%

systemic pressure, and pulmonary vascular resistance is less than one third of systemic resistance.¹⁰

Even if PAPVC is misleadingly diagnosed, there is still a chance to salvage when the drainage site of the pulmonary vein is routinely checked during ASD surgery. However, when transcatheter treatment is performed and PVPAC is omitted, it will be disastrous, as described in this case. This highlights the significance of confirming PAPVC in ASD surgery.

Patients with ACHD should undergo TTE for initial assessment, with timing of serial assessment based on anatomic and physiological severity and clinical status.¹⁰ In the preoperative preparation, it is necessary to find an experienced ultrasound physician to do preoperative ultrasound examination.¹¹ It has been reported that TTE using 2-dimensional color flow Doppler imaging and 2-dimensional is very helpful in the diagnosis of PAPVC in children. However, it may not apply to adults.² In patients with PAPVC, the origin, drainage site, and size of the abnormal pulmonary vein are variants. Moreover, the anatomic location of the left atrium and pulmonary veins is behind the heart. Therefore, for TTE, it is difficult to clearly position all the pulmonary veins. In addition, some patients are unable to get satisfactory images because of emphysema, obesity, chest wall deformities, and obstruction of ribs.

TTE has limited use in the assessment of anomalous pulmonary venous connections in adults with ASD; however, transesophageal echocardiography (TEE) is excellent for visualization of the entire atrial septum and pulmonary venous connections.¹⁰ Although it has been reported that the accuracy of TEE in the diagnosis of PAPVC is higher than that of transthoracic echocardiography, there is still great challenges in the diagnosis of PAPVC.¹²

Echocardiographic evaluation before device ASD closure aims to verify pulmonary veins connection and to assess defect rims. The disproportion between the right ventricle enlargement and the size of the ASD should alert the sonographer of a possible associated PAPVC. A complementary evaluation should be carried out before ASD closure, either by a cardiac scanner or by a TEE at the time of the intervention (ASD closure is usually guided by TEE). Pulmonary angiography also made it possible to verify the pulmonary venous connection before the implantation of the device. In adults with isolated secundum ASD causing impaired functional capacity, right atrial and/or RV enlargement, and net left-to-right shunt sufficiently large to cause physiological sequelae without cyanosis at rest or during exercise, transcatheter or surgical closure to reduce RV volume and improve exercise tolerance is recommended, provided that systolic PA pressure is less than 50% of systolic systemic pressure and pulmonary vascular resistance is less than one third of the systemic vascular resistance.¹⁰ Primum, sinus venosus, and coronary sinus ASDs should be closed surgically because

of the absence of appropriate rims for percutaneous device placement and the proximity of the atrioventricular valves and conduction system to the closure device.¹⁰ Criteria for attempted transcatheter TTE-guided closure included single secundum ASD, maximum defect diameter <38 mm in all views, and septal rims, with the exception of the anterior-superior rim, of ≥ 5 mm.¹³

In addition to echocardiography, computed tomography (CT) and cardiovascular magnetic resonance imaging (CV-MRI) should also be considered for diagnosis. However, CT has ionizing radiation and carcinogenic effects.¹⁴ Therefore, CV-MRI can be used as an alternative method for diagnosing PAPVC. In recent years, high-definition images of systemic and pulmonary veins can be obtained through CV-MRI, as well as accurate quantification of QP/QS ratio and dynamic images of the heart. More importantly, in the evaluation of right ventricular function and volume, CV-MRI is more accurate than other diagnostic modalities. Therefore, CV-MRI has been widely accepted in cardiac surgery. On the other hand, it is noisy and time-consuming to refer for MRI, so children may need to be sedated.¹⁵ As CV-MRI or CT is recommended for the evaluation of PAPVC,¹⁰ TTE and CTA were performed preoperatively in this case.

Although great progress has been made in the diagnosis and treatment of ASD, it is still possible to omit PAPVC. Interventional physicians must be familiar with the characteristics of various congenital heart diseases and select appropriate examination before closing the defect. When the diagnosis is suspicious during the treatment, it is necessary to make a timely and comprehensive examination to get a definitive diagnosis in order to take the correct treatment.

4 | CONCLUSION

A comprehensive examination is mandatory in the diagnosis of ASD before treatment, including echocardiography, CV-MRI, and/or CT, to verify other congenital heart diseases.

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Published with written consent of the patient.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTION

All authors: were involved in the writing, revisions, and final review of the manuscript.

ETHICAL APPROVAL

Institutional review board approval for case report is not required at our institution. To keeping ethical principles, name

of the patient was not pointed in the paper and the right of the subject was protected. Written informed consent was obtained from the patient for the publication.

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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SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section.

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