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Case Report

Neglected Partial Anomalous Pulmonary Venous Return during Atrial Septal Defect Device Closure

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Abstract

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital heart malformation in which 1 or more pulmonary veins drain into the systemic venous circulation or directly into the right atrium instead of the left atrium. It may occur alongside other congenital heart defects, including atrial septal defect (ASD). All patients with newly diagnosed ASD must be evaluated thoroughly for the likelihood of PAPVR to select surgical or percutaneous procedures.

Here, we describe a 10-year-old girl with PAPVR who underwent percutaneous device closure of her secundum ASD with her PAPVR neglected at 3 years of age. We had to correct the anomalous venous connection by removing the device during an intricate procedure. The typical connection of the pulmonary veins to the left atrium was reported on her postoperative echocardiography 1 day after surgery. The patient was discharged without complications, and her first follow-up visit 7 days after discharge was unremarkable. While the most accurate diagnostic tools for PAPVR are cardiovascular magnetic resonance imaging and computed tomographic angiography, a careful examination of the pulmonary veins during pulmonary angiography or transesophageal echocardiography in children helps identify PAPVR in patients with ASD.

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Introduction

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital heart malformation that occurs in approximately 0.4% to 0.7% of the general population.1 In this anomaly,[1] or more pulmonary veins drain into the systemic venous circulation or directly into the right atrium in lieu of the left atrium,[1] resulting in a left-to-right shunt with a broad spectrum of presentations. However, PAPVR is often asymptomatic and goes undiagnosed. In some cases, this condition eventually leads to pulmonary arterial hypertension. Early surgical repair is the treatment of choice.[2]

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PAPVR can occur on either side or bilaterally, with the latter being rare.[2,3] In 80% to 90% of cases, PAPVR originates from the right lung with 1 or more pulmonary veins draining most commonly into the superior vena cava or the right atrium and less commonly into the inferior vena cava, the innominate vein, and the coronary sinus.[1,2,5,6] Left-sided PAPVR usually drains into the left brachiocephalic vein via an anomalous vertical vein.[2,3]

PAPVR may occur as an isolated anomaly or alongside other congenital heart defects.[1,3] About 10% to 15% of secundum ASD cases and 85% of sinus venosus ASD cases are associated with PAPVR.[1,5] While the percutaneous closure of secundum ASD is an effective and safe treatment modality, all patients with newly diagnosed ASD must be examined thoroughly for the presence of PAPVR before an attempted closure, whether surgical or percutaneous.[5,6] PAPVR is missed frequently in transthoracic echocardiography (TTE).[2] Thus, a comprehensive examination, including transesophageal echocardiography (TEE), cardiovascular magnetic resonance imaging (CMR), computed tomography (CT), or conventional angiography (during device closure), should be performed before percutaneous ASD closure.[1,5]

Here, we describe a 10-year-old girl with PAPVR who underwent percutaneous device closure of her secundum ASD with her PAPVR neglected at 3 years of age. In this complicated patient, the anomalous venous connection was surgically corrected through a very intricate procedure by removing the device and normalizing the conduction of the pulmonary veins.

Case Report

We received a 10-year-old girl complaining of fatigue and dyspnea on exertion with a history of secundum ASD occlusion with a 15 mm Occlutech Septal Occluder (Occlutech GmbH, Jena, Germany) at 3 years of age in another center. Anomalous pulmonary veins were not identified at the time of percutaneous ASD closure in the primary center despite contrast injection into the pulmonary artery. Considering the neglected PAPVR, right ventricular enlargement, and a shunt flow greater than 4 found on the patient's echocardiogram, she was admitted for diagnostic cardiac catheterization.

Diagnostic cardiac catheterization was performed under general anesthesia through the right femoral vein. The venous catheter entered all 3 right pulmonary veins from the right atrium, and partial anomalous drainage of all right pulmonary veins was shown upon right pulmonary artery branch injection (approximately >50% of the pulmonary circulation) (Figures 1). Left pulmonary artery injection showed normal drainage of the left pulmonary veins. No left superior vena cava was detected. The patient was referred to a pediatric cardiac surgeon for surgical repair.

Through a median sternotomy under general anesthesia, the heart was exposed. The right atrium, the right ventricle, and the pulmonary artery were significantly dilated on inspection. On complete cardiopulmonary bypass and after opening the right atrium, no sinus venosus ASD was found. Nonetheless, all the right pulmonary veins were directly connected to the right atrium, which confirmed PAPVR (Figures 2). The ASD device was in a good position. The device was surgically removed as no route existed for redirecting the right pulmonary veins to the left atrium. Device excision was performed meticulously, and a large ASD was established. The PAPVR was corrected by directing all the right pulmonary veins to the left atrium and closing the created ASD with a pericardial patch. The patient was transferred to the intensive care unit in good condition and with stable hemodynamics.

Echocardiography on the first postoperative day showed a typical connection between the pulmonary veins and the left atrium, no residual ASD, reduced right atrial size, and acceptable global myocardial function.

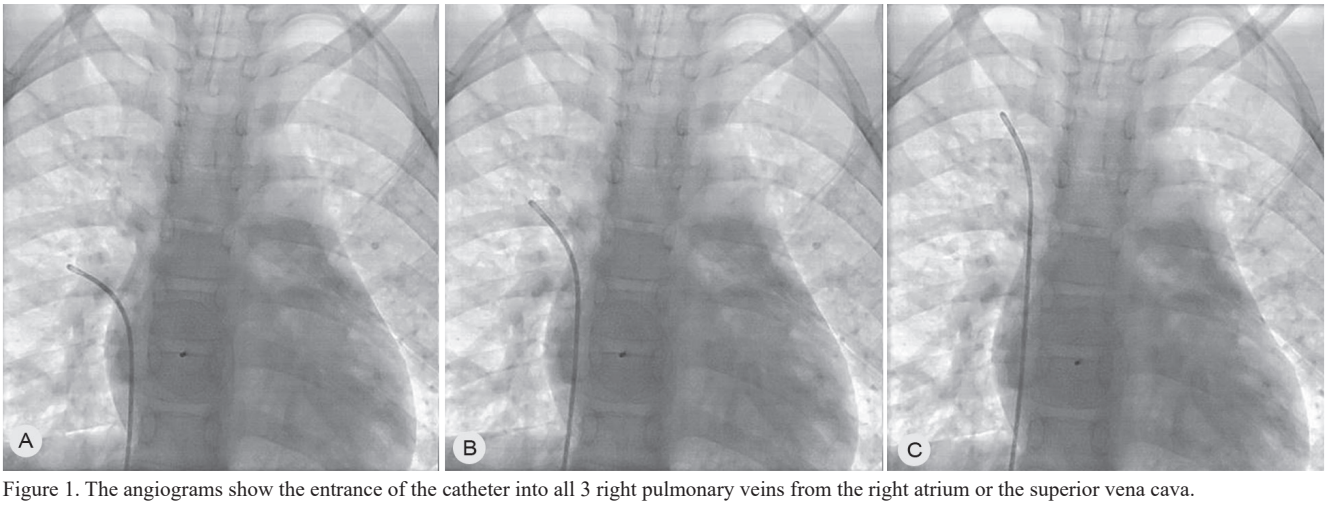

Figure 1. The angiograms show the entrance of the catheter into all 3 right pulmonary veins from the right atrium or the superior vena cava.
A to C) from the lowest to the highest

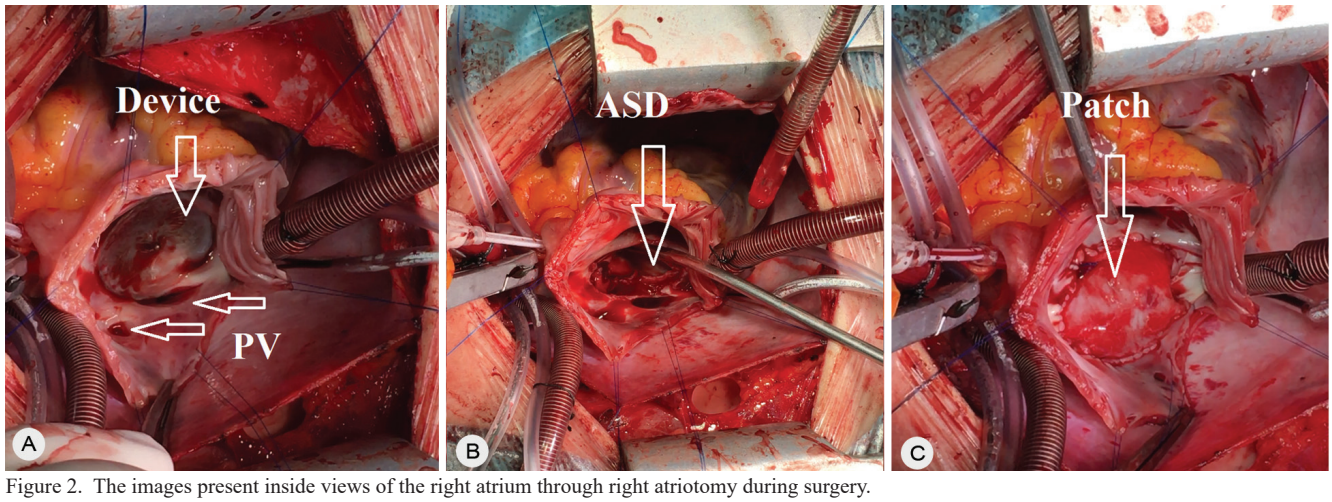

Figure 2. The images present inside views of the right atrium through right atriotomy during surgery.
A) The device (arrow) and the PV orifices opening into the right atrium (arrows)
B) The surgically created ASD after device removal (arrow)
C) The closing of the defect with a pericardial patch and the directing of the right PVs into the left atrium under the patch (arrow)
PV, Pulmonary vein; ASD, Atrial septal defect

The patient was transferred to the ward, where she had an uneventful postoperative period and was discharged without complications. Her follow-up echocardiography 7 days after discharge demonstrated no residual shunts, no obstruction to the pulmonary venous flow, and acceptable cardiac function.

Discussion

Surgery is the treatment of choice for patients with combined ASD and PAPVR. Nevertheless, in the following 2 situations, percutaneous ASD device closure is preferred, and the patient should not be deprived of its benefits: cases with a single small pulmonary vein with anomalous drainage (due to its negligible hemodynamic effects) and cases with a left-sided PAPVR who can undergo a hybrid procedure, with the ASD closed percutaneously and the PAPVR corrected surgically via a lateral thoracotomy without cardiopulmonary bypass.[7] Therefore, a thorough investigation for PAPVR is mandatory before any attempt at ASD closure.[1,5-8]

Despite the advances made in the diagnosis of ASD, formidable challenges still exist in the diagnosis of PAPVR.[1,9] TTE is not a reliable tool for identifying PAPVR in adults or children.[1,2,9] TEE, in contrast, can reveal PAPVR in children but is not accurate for the diagnosis of PAPVR in adults because it carries a high possibility of misdiagnosis.[1,5,6,8] Intracardiac echocardiography is not practical for the diagnosis of PAPVR either.[5,6] Overall, echocardiography alone does not suffice to establish a definite diagnosis of PAPVR.[1,8] A report by Al-Bustami et al is a fine example in that it describes an adult patient with ASD whose PAPVR remained undiagnosed despite 1 TEE and 2 TTE examinations before percutaneous ASD closure.[6]

Identifying PAPVR is possible during the levophase of pulmonary artery angiography if performed meticulously.[1,5,7] Still, the most accurate diagnostic tools for PAPVR are CMR and CT angiography with contrast injection.[1,4-6,10] CT exposes patients to ionizing radiation, which can be carcinogenic in children less than 6 years of age.[11] For children, it is preferable to perform a CMR instead of a CT scan.9 It should be noted that while CMR is easily obtainable in adults, performing it in children poses some limitations and can be challenging as it requires the child's cooperation or sedation.[1,10]

Conducting a CT scan or CMR is not common before percutaneous ASD closure, resulting in missed PAPVR in a small percentage of patients who will require surgical correction in the future. Consequently, such misdiagnoses should be avoided by performing CMR or CT in all patients with ASD before attempting a percutaneous ASD closure. Nonetheless, a careful examination of the pulmonary veins during pulmonary angiography or TEE in children can be beneficial in identifying PAPVR in almost all such cases.

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