

Cavopulmonary window: case report of an unusual variant of a sinus venosus defect

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Introduction

Isolated partial anomalous pulmonary venous return (PAPVR) with intact atrial septum is a rare finding. A cavopulmonary window is a side-to-side veno-venous communication of the right upper pulmonary vein with the superior vena cava which in its course retains connection to the left atrium.

Case presentation

We present a case of this unusual variant of a sinus venosus defect far from the atrial roof. Haemodynamic significance of the shunt was confirmed by enlargement of right heart cavities, elevation of pulmonary artery pressure, and significant left-to-right shunting using multimodality cardiac imaging (transoesophageal echocardiography, cardiac magnetic resonance imaging, and right heart catheterization). The defect has been successfully repaired using minimally invasive axillary thoracotomy.

Discussion

Partial anomalous pulmonary venous return prevalence is low and about 0.4–0.7% in autopsy series of patients with congenital heart disease. This patient's unusual variant of a sinus venosus defect with a window between a pulmonary vein and the superior vena cava far from the atrial roof shows that a sinus venosus defect is not a true atrial septum defect. Left-to-right shunting generally increases with age. Usually, surgical treatment is considered in cases of significant left-to-right shunt ($Q_p:Q_s > 1.5\text{--}2.0$) and right heart dilatation.

Keywords

Partial anomalous pulmonary venous return • Cavopulmonary window • Sinus venosus defect • Right cavity enlargement • Congenital • Case report

Learning points

- A cavopulmonary window may cause significant left-to-right shunting and can be corrected using minimally invasive surgery.
- A cavopulmonary window is an extreme form of a sinus venosus defect, which is not a true atrial septum defect.

Introduction

Partial anomalous pulmonary venous return (PAPVR) has first been mentioned by Winslow¹ in 1739 and fully described by Wilson² in 1798. The first *in vivo* diagnosis without exploratory thoracotomy is attributed to Dotter *et al.*³ in 1949 and was made by angiocardiography and cardiac catheterization.

Partial anomalous pulmonary venous return encompasses a specific group of congenital anomalies that are caused by the abnormal return of one or more, but not all, of the pulmonary veins directly or indirectly to the right atrium (RA). The vast majority of PAPVR is associated with a cardiac defect such as a secundum atrial septal or an upper sinus venosus defect.

In PAPV drainage pulmonary veins retain their normal connection to the left atrium (LA). Due to other intracardiac defects (such as a sinus venosus defect), oxygenated blood from the anomalous vein can enter the RA. Apart from patients with Turner syndrome⁴, isolated (intact atrial septum) PAPVR is a rare finding.

We report the case of a side-to-side veno-venous communication of the right upper pulmonary vein (RUPV) with the superior vena cava (SVC) which in its course retained connection to the LA (cavopulmonary window). The term cavopulmonary window should not

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be confused with cavopulmonary connection, a term usually used for surgically created connections from the big veins to the pulmonary arteries in single ventricle physiology.

Timeline

Medical history	Medical history significant for enlargement of right heart cavities with mild elevation of pulmonary artery pressure and mild dyspnoea. Electrocardiogram shows normal sinus rhythm with incomplete right bundle branch block, cardiac auscultation a fixed split of S2.
6 months prior to operation	<i>Transoesophageal echocardiography</i> shows partial reverse flow of the right upper pulmonary vein (RUPV) and evidence of relevant shunt at the level of the superior vena cava (SVC). <i>Cardiac magnetic resonance</i> imaging demonstrates a large side-to-side communication (cavopulmonary window) of the RUPV with SVC. The RUPV retains its connection to the left atrium (LA) in its course. Flow measurements show significant left-to-right shunt.
5 months prior to operation	<i>Right heart catheterization</i> shows a jump in the oxygen saturation of the SVC proximal to the cavopulmonary window and confirms significant left-to-right shunting.
Operation	An atrial tunnel patch is inserted through a patent foramen ovale to redirect flow from the RUPV to the LA using minimally invasive axillary thoracotomy.
7 months after operation	<i>Cardiac magnetic resonance</i> imaging demonstrates that the cavopulmonary window is closed with unchanged normal connection of the RUPV to the LA.

Case presentation

A 53-year-old female patient presented with a 10-year history of enlargement of right heart cavities with mild elevation of pulmonary artery pressure and mild dyspnoea. Her baseline electrocardiogram showed normal sinus rhythm with incomplete right bundle branch block, cardiac auscultation revealed a fixed split S2. First echocardiographic cue of the congenital disease was a partial reverse flow of the RUPV (Figure 1A, B) and evidence of relevant left-to-right shunt at the

level of the SVC (Figure 1C, D), as assessed by transoesophageal echocardiography. Her past medical history includes dyslipidaemia, arterial hypertension and anxiety disorder.

Cardiac magnetic resonance imaging (CMR) confirmed right heart dilatation (Figure 2A) with normal biventricular function (Figure 2B, visual assessment) and showed dilatation of the pulmonary trunk (38 mm). 3D magnetic resonance angiography (Golden-angle RAdial Sparse Parallel MRI) demonstrated a large side-to-side communication (cavopulmonary window) of the RUPV with SVC of 19 mm (Figure 3) resulting in isolated PAPVR. The RUPV retained its connection to the LA and as a result forming a PAPV drainage.

Flow measurements (Figure 4) were done of the ascending, and descending aorta, as well as pulmonary artery (not shown) and showed a significant left-to-right shunt with $Q_p:Q_s$ of 1.6. Approximate (no axial correction) net forward flow in the SVC distal to the cavopulmonary window was markedly elevated (113 mL). It was much higher than the usually expected backflow from head and upper extremities (difference of net forward flow of ascending and descending aorta: 96 mL - 63 mL = 33 mL) and further confirmed significant left-to-right shunting.

At right heart catheterization, the anomalous connection of the RUPV to the SVC was confirmed by a jump in the oxygen saturation of the SVC proximal to the cavopulmonary window of 76–96% at the level of the cavopulmonary window. Angiographically, there was evidence of significant left-to-right shunting with a $Q_p:Q_s$ of 2.2.

Given the haemodynamic significance of this patient's shunt, right cavity dilatation, and mild elevation of pulmonary artery pressure the defect has been repaired using minimally invasive axillary thoracotomy (Figure 5A). An atrial tunnel patch was inserted through a patent foramen ovale (PFO) to redirect flow from the RUPV to the LA (Figure 5B).

In a follow-up CMR 7 months after the operation, it could be demonstrated that the cavopulmonary window was closed with unchanged normal connection of the RUPV to the LA. Yet, there was evidence of a post-operative interatrial shunt ($Q_p:Q_s = 1.2$) in proximity to the ostium of the inferior vena cava. Right ventricular end-diastolic volume (RV EDV) was in the upper normal range (RV EDV indexed 106 mL/m²).

Discussion

Partial anomalous pulmonary venous return prevalence is low and about 0.4–0.7% in autopsy series of patients with congenital heart disease.⁵ More than 90% of patients with PAPVR draining into the SVC or the RA have an associated atrial septum defect (ASD) or sinus venosus defect.⁶ In the absence of associated anomalies, patients often remain asymptomatic, thus explaining the difficulty in knowing the real incidence of isolated and/or asymptomatic PAPVR.¹

A sinus venosus defect has been defined as an anomalous pulmonary vein communication to a systemic vein with an interatrial

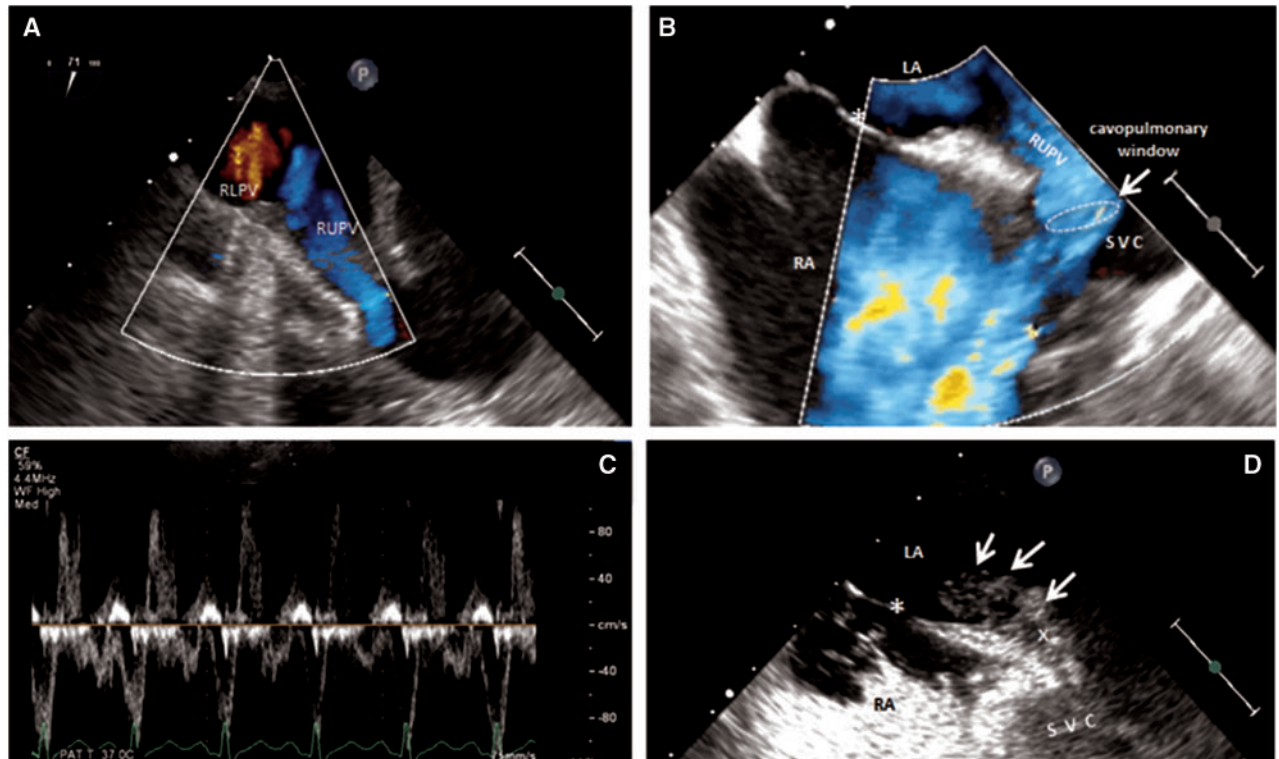


Figure 1 (A) Mid-oesophageal view. Late diastolic still frame at 71° . Colour flow of the right pulmonary veins shows a retrograde flow (blue) in the right upper pulmonary vein and an antegrade flow (red) in the right lower pulmonary vein. (B) Additional pulsed-wave Doppler demonstrates a pendular flow (alternately antegrade and retrograde) in the right upper pulmonary vein. (C) Mid-oesophageal view. End-diastolic still frame at 135° . Colour flow shows the atrial left-to-right shunt over the right upper pulmonary vein via cavopulmonary window and superior caval vein. *Interatrial septum. (D) Mid-oesophageal view. Still frame at 127° after injection of agitated colloid into the right cubital vein. The contrast agent predominantly reaches the right atrium via superior caval vein. Some contrast, however, enters the left atrium over passage through the right upper pulmonary vein (arrows). *Interatrial septum, xright upper pulmonary vein. RUPV, right upper pulmonary vein; RLPV, right lower pulmonary vein; RA, right atrium; LA, left atrium; SVC, superior vena cava.

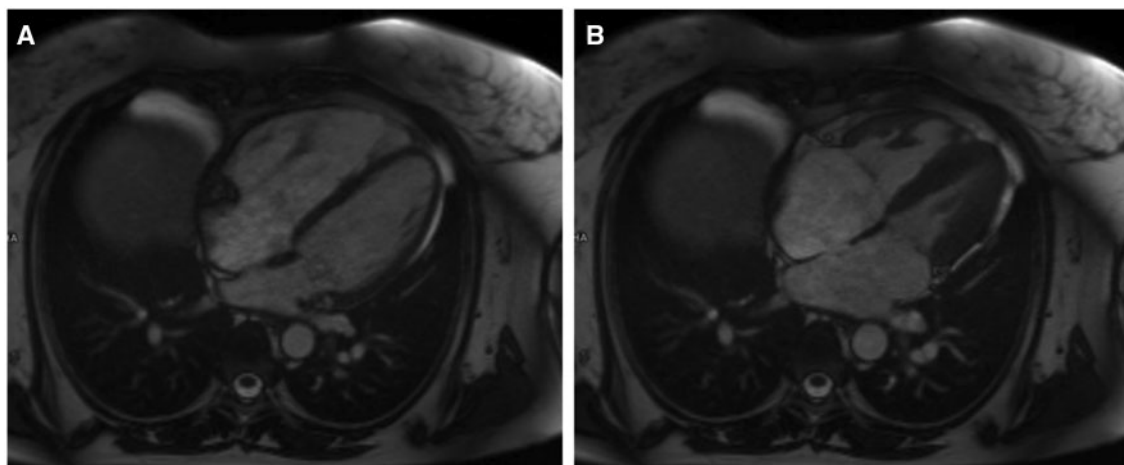


Figure 2 (A) End-diastolic frame of a cine balanced steady state free precession sequence showing right ventricular dilatation (visual assessment). (B) End-systolic frame of a cine balanced steady state free precession sequence showing normal right ventricular function (visual assessment) and atrial dilatation.

communication outside the confines of the atrial septum, in the unfolding wall that normally separates the LA from either caval vein. It results in overriding of the caval veins across the intact atrial septum and partial pulmonary vein anomalous drainage.^{7, 8} This patient's unusual variant of a sinus venosus defect with a window between a pulmonary vein and the superior vena cava far from the atrial roof shows that a sinus venosus defect is not a true ASD.⁷ Embryologically, Butts *et al.*⁹ have outlined that so-called sinus

venosus defect is the consequence of persistence of foetal systemic to pulmonary veno-venous bridges, rather than of deficiencies in atrial septation. Pathophysiologically, a cavopulmonary window leads to right heart volume overload due to recirculation of already oxygenated pulmonary venous blood through the pulmonary vasculature (left-to-right shunting).

Left-to-right shunting generally increases with age. Usually, surgical treatment is considered in cases of significant left-to-right shunt ($Q_p:Q_s > 1.5-2.0$) and right heart dilatation. Flow measurements—both invasive and non-invasive—are error prone and should always be evaluated in the further imaging (chamber dilatation) and clinical context.

Unlike in the case report of Aramendi *et al.*,¹⁰ this case report demonstrates a different surgical approach with an atrial tunnel patch that was inserted through a PFO to redirect flow from the RUPV to the LA.

Conclusion

A cavopulmonary window results in normal pulmonary venous connection (RUPV → LA) with partially abnormal veno-venous drainage (RUPV ↔ SVC) which can be haemodynamically significant.

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Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

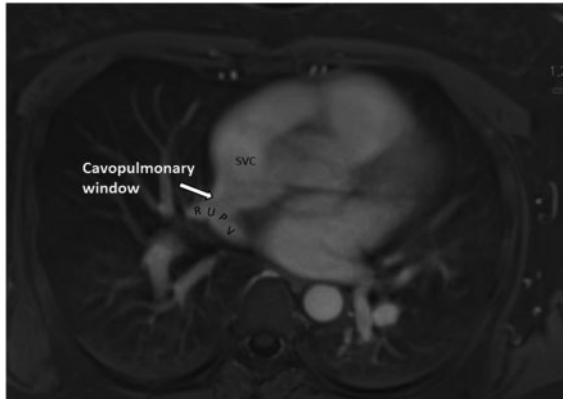


Figure 3 3D magnetic resonance angiography (GRASP) showing a 19 mm side-to-side communication of the superior vena cava and the right upper pulmonary vein due to unroofing of the wall that normally separates the right upper pulmonary vein from the superior vena cava. The right upper pulmonary vein retains connection to the left atrium in its course (*cavopulmonary window*, see also *Figure 1B*). The connection of the right upper pulmonary vein with the superior vena cava is at the ostium of the superior vena cava with the right atrium. GRASP, Golden-angle RADial Sparse Parallel MRI.

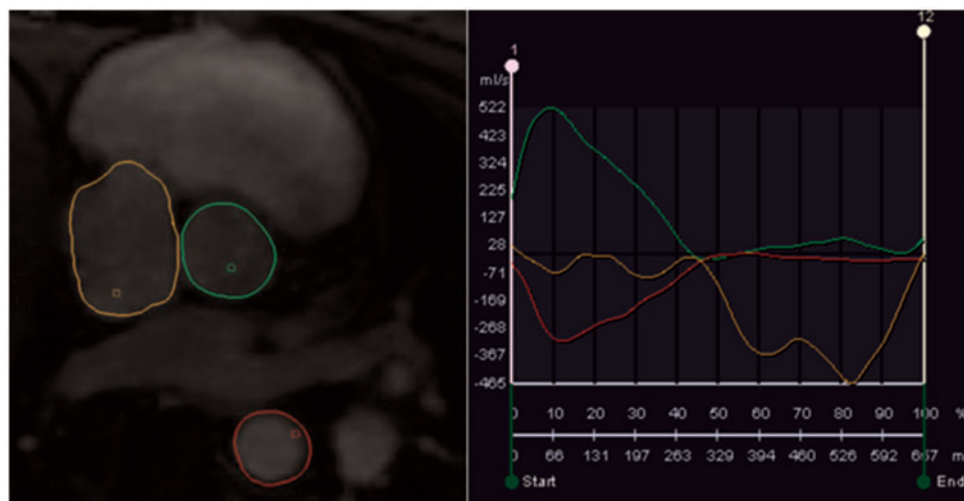


Figure 4 Flow measurements of ascending aorta (green encircling), descending aorta (red), and approximate (not axis-correct) measurement of superior vena cava distal to the cavopulmonary window (orange), as well as pulmonary artery (not shown).

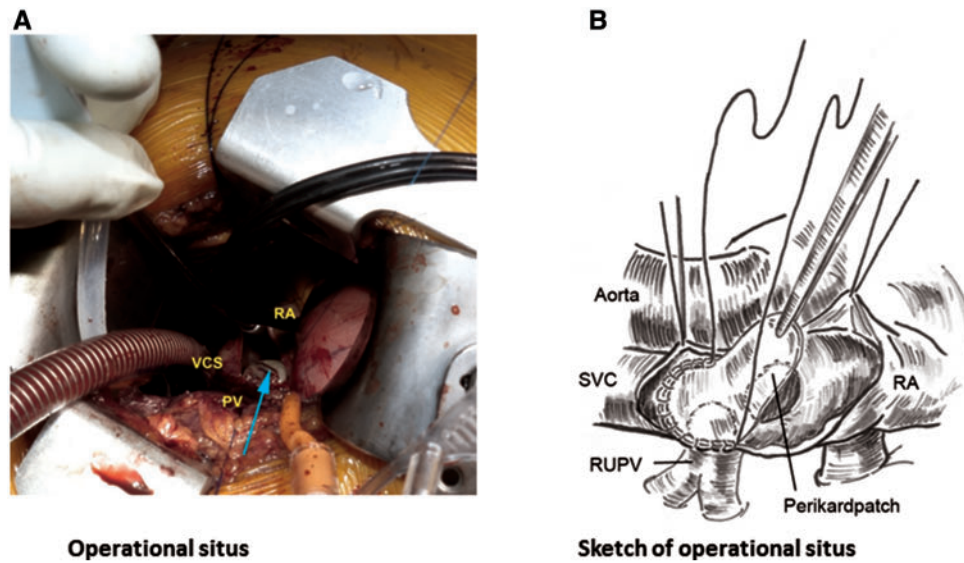


Figure 5 (A) Intraoperatively, the cavopulmonary veno-venous window could be confirmed 1 cm far from the connection of the superior vena cava with the right atrial roof. An atrial tunnel patch through a patent foramen ovale was inserted to redirect flow from the right upper pulmonary vein to the left atrium by minimally invasive axillary thoracotomy (B). Atrial tunnel patch to redirect flow from the right upper pulmonary vein to the left atrium.

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