

CLINICAL VIGNETTE

Unusual Variant of Partial Anomalous Pulmonary Venous Connection to Coronary Sinus

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Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital cardiovascular disorder characterized by some, but not all, of the pulmonary veins connecting to the systemic venous system. This connection results in oxygenated blood draining directly or indirectly into the right atrium. In patients with PAPVC, a large degree of left to right shunting often results in symptoms associated with pulmonary over-circulation and right heart failure requiring surgical repair in most cases. While most PAPVC cases report pulmonary veins draining into the right atrium in association with a sinus venous defect, the left innominate vein, superior vena cava, or inferior vena cava (Scimitar syndrome), few cases of PAPVC with the pulmonary vein draining into the coronary sinus have ever been reported.

Case Report

A 37-year-old female with no past medical history presented with progressive dyspnea on exertion and bilateral leg swelling over a few months.

Physical exam was significant for a 2/6 systolic murmur noted at the right upper sternal border and trace bilateral pitting edema. EKG showed sinus rhythm with no evidence of right atrial or ventricular abnormality. CXR was normal. On trans-thoracic echocardiogram (Figures 1-2), there was evidence of right atrial and ventricular dilatation with septal flattening suggestive of volume overload. Mild pulmonary hypertension with RV/PASP, estimated at 44 mmHg, and a dilated coronary sinus were also noted. Saline contrast study was negative for atrial septal defects, and there was no significant tricuspid or pulmonic regurgitation. Trans-esophageal echocardiogram was performed to assess for possible atrial level defect and was suspicious for anomalous connection of the left upper pulmonary vein to the coronary sinus. Cardiac CT (Figures 3-5) confirmed the diagnosis of PAPVC with both left upper and lower pulmonary veins draining into the coronary sinus.

The patient subsequently underwent surgical repair, which included pericardial patch closure of the coronary sinus orifice and creation of a window between the coronary sinus and left atrium redirecting anomalous pulmonary vein flow and

attenuating the left to right shunt. The patient tolerated the procedure well with subsequent improvement of symptoms.

Discussion

PAPVC is a rare congenital disorder with overall incidence at autopsy estimated at 0.6-0.7%.¹ PAPVC more commonly involves the right pulmonary veins with only 10% involving the left pulmonary veins.² PAPVC is usually associated with atrial septal defect with few cases ever reported with an intact atrial septum.³ Also, few PAPVC case reports were found in which the left pulmonary vein drains into the coronary sinus.⁴ In our case, the patient had an abnormal connection between both the upper and lower pulmonary veins, connecting to the coronary sinus with an intact atrial septum. To our knowledge, this anatomic variant of PAPVC has not been reported in the literature.

Cases of isolated PAPVC are usually reported as incidental findings, and many patients remain asymptomatic.⁵ Patients with symptoms typically present with dyspnea on exertion and fatigue that may progress to heart failure if the degree of left to right shunting is large. Physical exam findings are also directly related to the degree of left to right shunting, which may present as precordial bulge or systolic ejection murmur heard along the left sternal border. Chest x-ray findings are nonspecific and may include cardiomegaly or increased pulmonary vascular markings.

Trans-thoracic echocardiogram may reveal right atrial and ventricular volume overload, manifested as chamber dilation and septal flattening. Once right atrial or ventricular dilation is recognized, additional evaluation is needed to determine the etiology. This includes assessment for significant tricuspid or pulmonic regurgitation, and a saline contrast study to evaluate for an atrial level shunt. Trans-esophageal echocardiogram and/or cardiac CT should be considered to assess for atrial septal defects and more rare causes of right atrial and ventricular enlargement, such as PAPVC.^{6,7}

Surgery is the best option for management of patients with PAPVC. The timing of surgery is currently controversial. Some believe early repair before the patient develops

symptoms of right heart volume overload and pulmonary hypertension is appropriate. Others believe surgery is indicated only if the patient becomes symptomatic as complications associated with surgical repair, such as atrial fibrillation, complete heart block, thus possibly leading to infection. In patients who undergo surgery, the outcome is generally excellent.⁸

Conclusion

Isolated partial anomalous venous connection is a rare finding. To our knowledge, this is one of the first case reports of a patient with anomalous pulmonary venous connection between the left upper and lower pulmonary veins to the coronary sinus without an atrial septal defect. In symptomatic patients with unexplained right atrial or ventricular dilatation, PAPVC should be included in the differential diagnosis and further diagnostic testing with a trans-esophageal echocardiogram and/or cardiac CT may be needed for complete assessment.

Figures

Figure 1: Transthoracic echocardiogram, apical four chamber view. Severe dilation of the right atrium and ventricle. RV = right ventricle, RA = right atrium, LV = left ventricle, and LA = left atrium.



Figure 2: Transthoracic echocardiogram, parasternal long axis view. Severe dilation of the coronary sinus. RV = right ventricle, LV = left ventricle, Ao = aorta, LA = left atrium, and CS = coronary sinus.



Figure 3: Cardiac CT axial view. Severe dilation of right ventricle, right atrium and coronary sinus. RV = right ventricle, LV = left ventricle, RA = right atrium, and CS = coronary sinus.

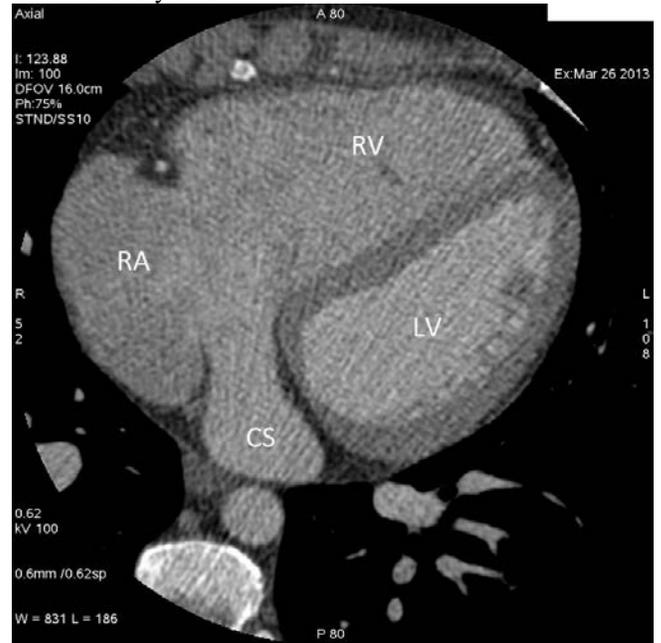


Figure 4: Cardiac CT coronal oblique view. Left lower pulmonary vein draining into coronary sinus. LLPV = left lower pulmonary vein, LA = left atrium, CS = coronary sinus, RA = right atrium, and SVC = superior vena cava.

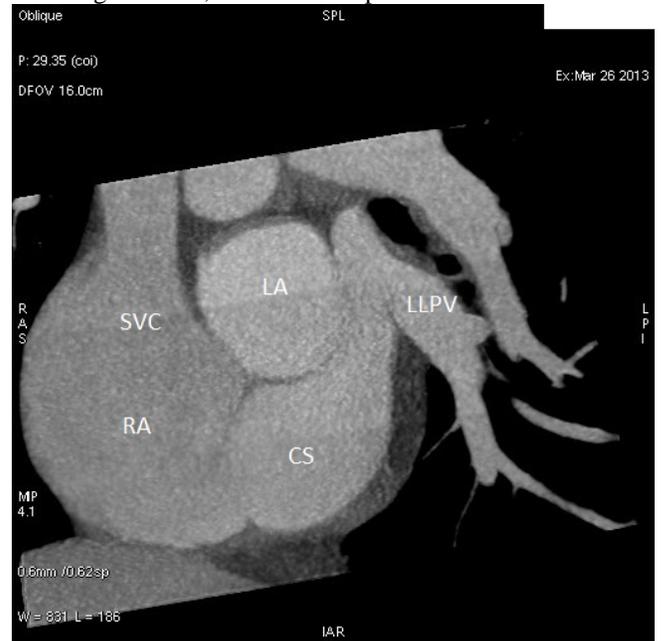
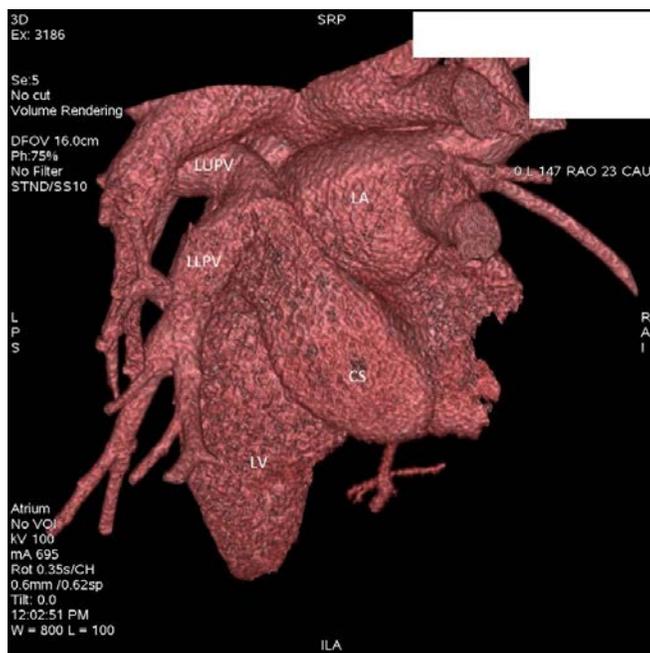


Figure 5: Volume rendered image of cardiac CT. Left upper and lower pulmonary veins seen draining directly into the coronary sinus. LUPV = left upper pulmonary vein LLPV = left lower pulmonary vein, LA = left atrium, CS = coronary sinus, and LV = left ventricle.



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