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Editorial

An unusual type of partial anomalous pulmonary venous return with all pulmonary veins draining to left atrium (!)

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A 66-year-old woman presented to the cardiology clinic with dyspnea and decreased exercise tolerance. Physical examination was significant for a precordial 2/6 systolic murmur. Vital signs were in normal range. ECG showed sinus rhythm with right axis deviation and right bundle branch block. Chest X ray was normal.

Transthoracic echocardiography (TTE) revealed right ventricular and right atrial dilatation, severe tricuspid regurgitation with normal tricuspid valve morphology, mild pulmonary arterial hypertension with an estimated pulmonary artery systolic pressure of 39 mm Hg and a normal left ventricular function. These findings brought to mind the possibility of left-to-right shunt and TEE was planned. On TEE examination all pulmonary veins were shown to drain into the left atrium and interatrial septum was found to be intact.

The discrepancy between the normal tricuspid valvular morphology and severe tricuspid regurgitation with right ventricular dilatation led to the consideration of unusual type of left-to-right shunt. CMR was performed for further information and verified that there was no atrial septal defect (ASD) and all pulmonary veins drained into the left atrium. In addition to these findings CMR revealed the presence of a connection between right upper pulmonary vein (RUPV) and superior vena cava (SVC). In its course of RUPV, it was connected to SVC at first, then reformed its structure again, eventually drained into the left atrium. (Figs. 1, 2, 3, Video 1).

The patient was referred to cardiothoracic surgery for surgical correction of the partial anomalous pulmonary venous return (PAPVR) and tricuspid repair.

PAPVR is a rare congenital cardiac pathology which should be suspected in case of unexplained right ventricular dilatation. PAPVR occurs where one or more, but not all, pulmonary veins drain into a systemic vein or the right atrium (RA) rather than the left atrium. Anomalous pulmonary veins may drain to the SVC, RA, inferior vena cava, azygos vein, brachiocephalic vein, hepatic vein, portal vein, innominate vein, coronary sinus, and hemiazygos vein [1,2].

Most commonly an anomalous RUPV drains to the SVC (74%) [3]. Interestingly, Ho et al. demonstrated that only half of the PAPVR were right sided which may be an indication that the pediatric and adult populations with PAPVR may be significantly different [4]. An associated ASD of the sinus venosus type is found 80–90% of cases of PAPVR; rarely is the atrial septum found to be intact [5].

PAPVR is often clinically silent, and found it in 0.4% of postmortem examinations [6]. It may present in the adult population and generally diagnosed in the work-up of a patient with exertional dyspnea and right ventricular dilatation. The defect becomes clinically overt if the return of anomalous pulmonary venous flow to the right side of heart severe enough. Persistent systemic venous connection acts as a left-to-right shunt, where the right ventricle dilates due to the volume overload. This leads to gradual functional tricuspid regurgitation. Over time, the increase in pulmonary blood flow can lead to pulmonary arterial hypertension [7].

PAPVR has a pathology of embryological origin in which failure of one or more of the main pulmonary vascular beds to connect with the common pulmonary vein, and maintain the persistent systemic venous connection results in PAPVR [8].

Noninvasive imaging and diagnosis of PAPVR continues to evolve. TTE, which is often the first cardiac imaging modality obtained in patients with dyspnea, cannot reliably delineate pulmonary venous anatomy. PAPVR is usually diagnosed by TEE but occasionally TEE may be inconclusive due to technical limitations or unusual anatomic variations as presented by our case. CMR is increasingly being used as it provides an accurate anatomic definition of the heart and vascular structures and evaluate the systemic veins, as well as the number, origin, course, and drainage of all pulmonary veins. It also quantifies the shunt volume accurately [9]. CMR may be specifically recommended as a complementary to TEE in any patient with an unexplained dilatation of the right sided heart chambers.

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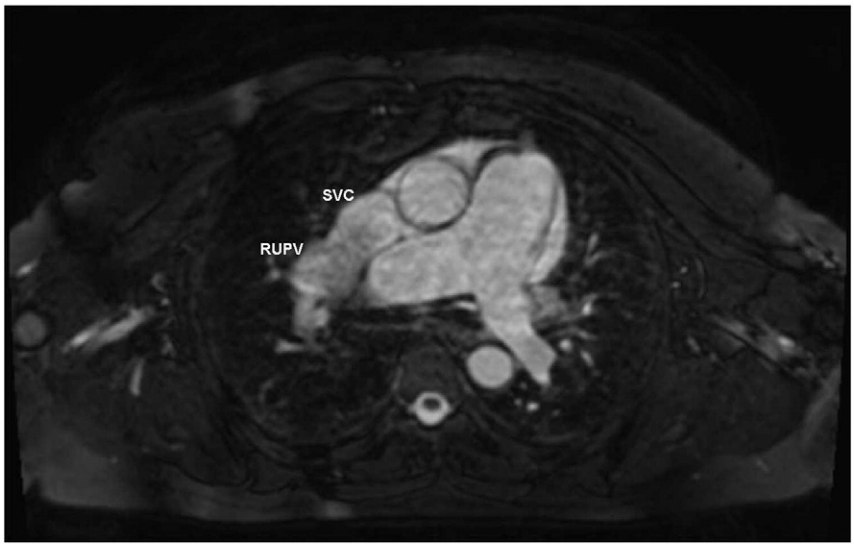

Fig. 1. Demonstrates the connection between RUPV and SVC.

Multidetector computed tomography (MDCT) scan is also effective diagnostic modality defining ASDs and PAPVR. Disadvantages of computed tomography include an inability to calculate shunt fraction, ionizing radiation, and a greater risk of nephrotoxicity with iodinated contrast than CMR with gadolinium [10].

The unusual connection between pulmonary veins and systemic venous system may remain undiagnosed if only TEE examination is obtained. As demonstrated by the cases discussed, although all pulmonary veins were shown to drain into the left atrium and interatrial septum was found to be intact by TEE, CMR revealed the connection between RUPV and SVC leading to left to right shunt and explained the cause of right ventricular dilatation. To the best of our knowledge there has been no report of a case in the literature describing the right upper pulmonary vein connecting with SVC firstly, then reforming its own structure and draining into left atrium.

Surgery is generally effective at correcting the abnormal shunt. Patients with significant right ventricular dilatation, as the cases discussed, need surgery. However, in asymptomatic patients with a low shunt fraction and no clinical or echocardiographic evidence of right ventricular overload, surgery may be unnecessary [11]. Unfortunately, in patients with irreversible severe pulmonary arterial hypertension, surgery is unlikely to change prognosis of disease and heart-lung transplant may be the only curative option. In these patients, medical therapy such as bosentan and phosphodiesterase inhibitors may cause some improvements in symptoms [12].

As a conclusion we describe a case of late clinical presentation of an unusual type of left-to-right shunt with intact atrial septum and all pulmonary veins were shown to drain into left atrium. To the best of our knowledge, this anatomic variation is the first report of the RUPV connecting with SVC firstly, then reforming its own structure and draining into left atrium. PAPVR is a rare cause of left to right shunt and may enlarge the right sided heart chambers. There should be a high index of suspicion of left-to-right shunt in patient with right ventricular dilatation and severe tricuspid regurgitation with normal tricuspid valve morphology. In routine clinical practice TEE is the modality of choice to diagnose this entity. But for unusual types of PAPVR, CMR is needed to detect the connection between pulmonary drainage and systemic venous system, establishing the cause of right ventricular dilatation.

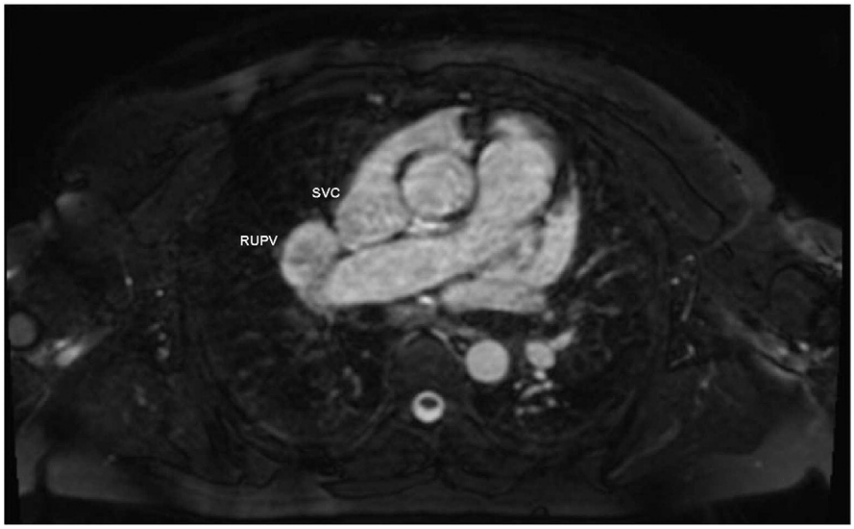

Fig. 2. Shows the reformed RUPV.

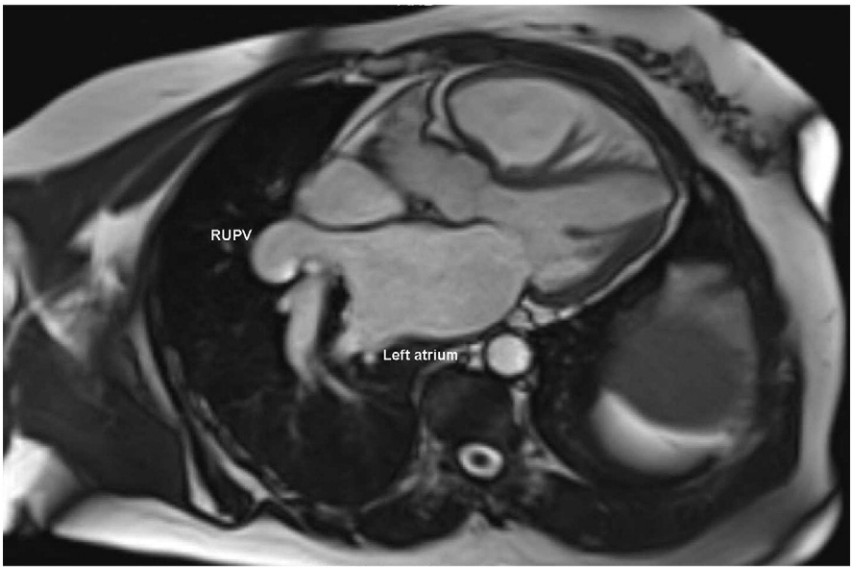

Fig. 3. Demonstrates the drainage of RUPV into the left atrium.

Supplementary data to this article can be found online at http://dx.doi.org/10.1016/j.ijcard.2016.08.055.

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

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