

A Rare Variant of Complex Partial Anomalous Pulmonary Venous Connections

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ABSTRACT

We report a 10 year old girl who was detected to have a sinus venosus atrial septal defect with complex partial pulmonary venous connections. Left pulmonary veins were draining into right superior vena cava via the innominate and vertical vein, and right upper pulmonary vein was draining directly into superior vena cava. Only the right lower pulmonary vein was connected to the left atrium. Though any combinations of pulmonary venous drainage are reported in literature, current reported type of pulmonary venous connections is one of the rarest type with only few cases previously reported. Through this report we also want to emphasize the need for screening for all pulmonary veins every case of atrial septal defect especially in sinus venosus type defect and when there is discrepancy between size of atrial septal defect and haemodynamic disturbance.

Key words: Atrial septal defect, Pulmonary venous connections, Sinus venosus.

BACKGROUND

Partial anomalous pulmonary venous connection (PAPVC) are rare group of congenital cardiac defects characterised by the abnormal return of one or more (but not all) of the pulmonary veins directly to the right atrium or indirectly through a variety of venous connections from the anomalous pulmonary vein. PAPVC is relatively a rare clinical condition with estimated prevalence of 0.7% in general population. We are reporting this case as this is one of the rarest variant of pulmonary venous connections and to highlight the importance of screening of all pulmonary veins in every case of atrial septal defect.

CASE REPORT

10 year old girl presented with a history of dyspnea on exertion of NYHA (*New York Heart Association*) class 2 since early childhood. There was no history of cyanosis or congestive cardiac failure. On examination there was no evidence of cyanosis or clubbing and vitals were within normal limits. Cardiovascular examination findings were suggestive of atrial septal defect with wide and fixed second heart sound with grade 3 ejection systolic murmur in left 2nd intercostal space. Baseline oxygen saturation was 94 % in all 4 limbs. Baseline electrocardiography revealed RsR pattern in right precordial leads with normal P wave axis. Chest Xray showed cardiomegaly with normal pulmonary vasculature with prominent left upper mediastinal shadow. 2D echocardiography with color doppler showed a 5 mm sinus venosus ASD with right ventricular and right atrial dilatation indicating a hemodynamically significant left to right shunt (figure 1A and 1B). Echocardiographically calculated left to right shunt fraction was 2:1 and right ventricular systolic pressure was 30 mm of Hg. Right lower

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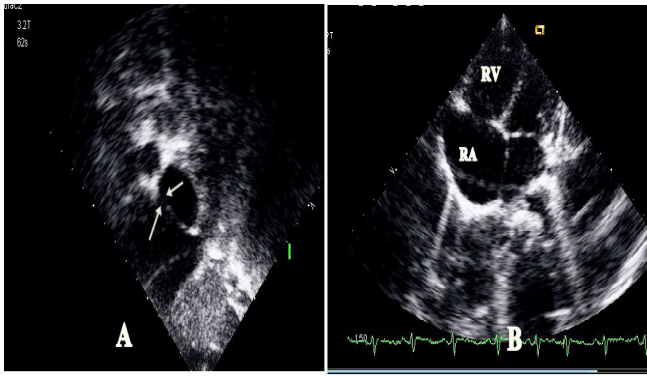


Figure 1: A-2D Echocardiography with subcostal bicaval view showing 5 mm sinus venosus atrial septal defect [arrow mark]. (SV ASD- sinus venosus atrial septal defect), B- apical 4 chamber views showing right atrial (RA) and right ventricular (RV) dilatation

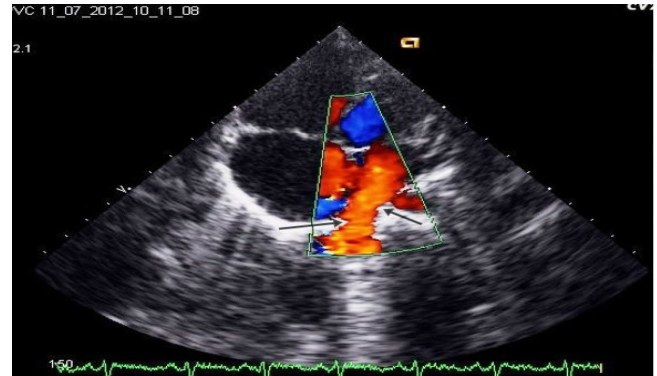


Figure 2: 2D Echocardiography with apical 4 chamber view showing right lower pulmonary vein draining into left atrium [arrow marks]



Figure 3: 2 D Echocardiography with suprasternal view demonstrated left sided pulmonary veins draining to innominate vein via the vertical vein (SVC-superior vena cava, VV-vertical vein)



Figure 4: Contrast enhanced CT scan showing right upper pulmonary vein draining into superior vena cava

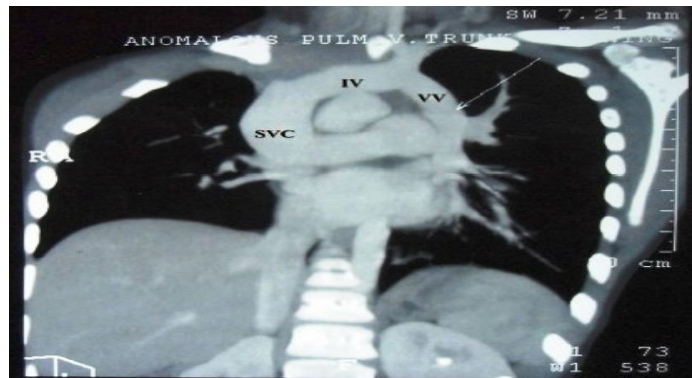


Figure 5: Contrast enhanced CT scan showing left pulmonary veins draining into superior vena cava via the innominate and vertical veins (SVC-superior vena cava, VV-vertical vein, IV-innominate vein)

pulmonary vein was normally draining into left atrium (Figure 2 and video 1) but right upper pulmonary vein and left pulmonary veins drainage into left atrium could not be delineated. Suprasternal view demonstrated the left sided pulmonary veins draining to innominate vein via the vertical vein (Figure 3 and video 2). But right upper pulmonary vein could not be identified in echocardiography. Contrast enhanced CT scan was done for the confirmation

of the diagnosis. CT thorax showed normal right lower pulmonary vein drainage. Right upper pulmonary veins were seen draining into superior vena cava (figure 4) and left pulmonary veins were connected into innominate veins via the vertical vein (figure 5). There were no other cardiac or extra cardiac anomalies present. In total final diagnosis was sinus venosus atrial septal defect associated with complex PAPVC, with only one pulmonary veins draining normally

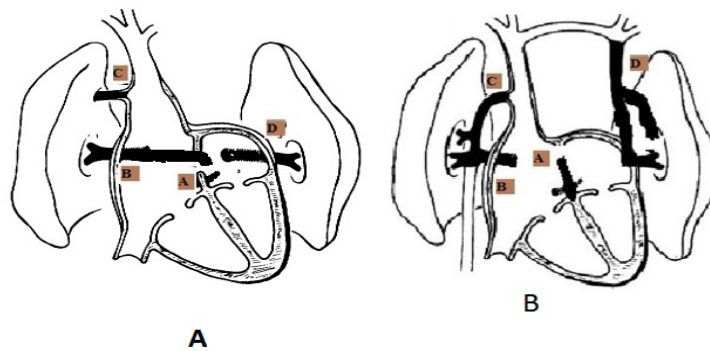


Figure 6: 6A- Schematic diagram showing the pulmonary venous connections of the most common type of partial pulmonary venous connection (A- Atrial septal defect, B- Right lower pulmonary veins draining into left atrium, C-Right upper pulmonary vein draining into superior vena cava, D-Left pulmonary veins draining left atrium). Figure 6B- Schematic diagram showing the pulmonary venous connections of the index reported case (A-Atrial septal defect, B- Right lower pulmonary veins draining into left atrium, C-Right upper pulmonary vein draining into superior vena cava, D-Left pulmonary veins draining into innominate veins via then vertical veins)

to left atrium. Patient was advised to undergo surgical correction of both atrial septal defect and the pulmonary venous drainage, but later was lost for follow-up.

DISCUSSION

Partial anomalous pulmonary venous return (PAPVC) is a rare entity with reported incidence of 0.4–0.7%. The most common presentation is a right upper lobe vein draining into either the right atrium or superior vena cava.¹ Right sided PAPVC are 10 times more common than left sided PAPVC. Left sided PAPVC represent only 10 % of the total cases.² PAPVC most commonly presents with atrial septal defect (ASD), reportedly in 80–90% of cases. Numerous variations of pulmonary venous drainage have been explained in literature. 2 of the largest published series of cases of abnormal pulmonary venous drainage by Snellen *et al* and Hu *et al* has reported 43 and 27 different variations of pulmonary drainage respectively.^{3,4} In our index case, both left pulmonary veins and right upper pulmonary vein was draining to right atrium as shown in this Schematic diagram (Figure 6). Such complex PAPVC has rarely been reported previously. In case series by Snellen *et al*, out of 124 cases only 2 cases has similar pulmonary venous drainage.³ In case series by Hu *et al*, out of 113 patients only 2 patients had similar pulmonary venous connections.⁴ Chest ray findings of partial pulmonary venous anomalies are similar to atrial septal defect. But in case of left sided TAPVC, prominent left sided vertical vein shadow is often seen. Differential considerations of an enlarged vertical vein include an enlarged left superior intercostal vein and persistent left superior vena cava which can be easily differentiated in echocardiography and CT scan.⁵ In earlier times, catheter-based angiography was the imaging modality of choice for confirmation of the

diagnosis; this has been superseded by echocardiography, computerized tomographic angiography, and magnetic resonance imaging. In our index case CT scan not only helped in diagnosis of the condition but also delineated the individual pulmonary veins and their connections. Long-standing PAPVC can predisposes the patient to right-sided volume overload, tricuspid regurgitation (TR), pulmonary hypertension, irreversible pulmonary vascular disease, right ventricular dysfunction. Current indications for the surgery are symptomatic PAPVC and asymptomatic patients with Qp:Qs >1.5, right ventricular dilation, pulmonary artery hypertension in order to prevent the development and progression to irreversible pulmonary vascular Disease.⁶ Our index reported case was referred for surgical correction in view of symptoms and significant left to right shunt. In view of the rarity of such complex pulmonary venous drainage pattern and only few reported cases, we planned to publish the case. We also want emphasize the importance of imaging for the individual pulmonary veins in every case of ASD, especially if hemodynamic Over load is disproportionate to the size of the ASD.

LEARNING POINTS/TAKE HOME MESSAGES

1. Screening for all pulmonary veins are an important exercise in every case of atrial septal defect especially in sinus venosus type defect and when there is discrepancy between size of atrial septal defect and haemodynamic disturbance.
2. Numerous combinations of pulmonary venous connections have been reported in Literature, ranging from abnormal connections of one to vein to all 4 veins.
3. Computerized tomographic angiography, and magnetic resonance imaging have Superseded echocardiography and conventional angiogram for imaging for pulmonary veins.

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