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CASE REPORT

A rare malformation of bilateral superior vena cava with bilateral partial anomalous pulmonary venous connection in the presence of ostium secundum atrial septal defect: management strategies and pitfalls

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Abstract Bilateral superior vena cava (SVC) with bilateral partial anomalous pulmonary venous connection is a very rare congenital cardiac malformation. Here, we are reporting a case of 18-year-old male who had bilateral SVC with bilateral anomalous pulmonary venous connection associated with ostium secundum atrial septal defect. The patient underwent successful surgical correction for the same.

Keywords Bilateral superior vena cava · Atrial septal defect · Partial anomalous pulmonary venous connection

Case details

The index case was an 18-year-old male, weighing 40 kgs, who presented with complaints of dyspnea on exertion NYHA class II since 3 months. The examination showed ejection systolic murmur with wide and fixed splitting second heart sound. Electrocardiographic examination showed RSR' pattern in lead V1. A chest radiograph showed the absence of cardiomegaly with increased pulmonary blood flow. Echocardiogram revealed ostium secundum atrial septal defect (ASD) with intact interventricular septum and moderate tricuspid regurgitation with mild pulmonary hypertension. Contrast-enhanced computed tomography confirmed the presence of ostium secundum atrial septal defect with bilateral superior vena cava (SVC), right SVC draining into right atrium and left SVC draining into right atrium through coronary sinus. A connecting vein was present between both cavae. It also showed right upper lobe pulmonary veins draining high into right SVC and left upper lobe pulmonary vein draining into left SVC. Left lingular and anteromedial basal segment veins were draining separately into left SVC. Right middle, lower lobe, and rest of left lower pulmonary veins were draining into the left atrium (Fig. 1).

The patient underwent surgical repair on cardiopulmonary bypass with moderate hypothermia and antegrade blood cardioplegia. The left SVC was ligated above pulmonary vein openings. Through a right atriotomy, right upper pulmonary vein baffled into left atrium using autologous glutarldehyde-treated pericardial patch. The coronary sinus was cutback, and the same patch was extended to close coronary sinus opening, leaving left upper pulmonary vein draining into left atrium through unroofed coronary sinus. Right SVC was repaired with autologous glutarldehyde-treated pericardial patch. Patient was rewarmed and was weaned off from cardio pulmonary bypass without any hemodynamic compromise or desaturation. The cardiopulmonary bypass time was 73 min, and aortic cross clamp time was 56 min. The post-operative course was uncomplicated, and the patient was discharged on the sixth post-operative day. The post-operative echocardiographic examination revealed all pulmonary veins draining into left atrium without any obstruction, no residual ASD with normal biventricular function. Post-operative computed topography revealed drainage of left upper pulmonary vein into left SVC, while all other pulmonary veins were draining into left atrium. The ligature of left SVC was just below the opening of left upper pulmonary vein (Fig. 2). Thus, we failed to divert left upper pulmonary vein. As the patient was asymptomatic with baseline saturation of 97% on room

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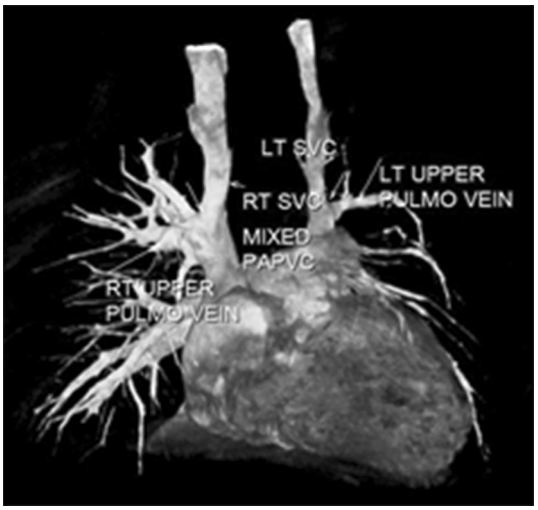

Fig. 1 Contrast-enhanced computed scan showing the presence of bilateral superior vena cavae with bilateral partial anomalous pulmonary venous connection

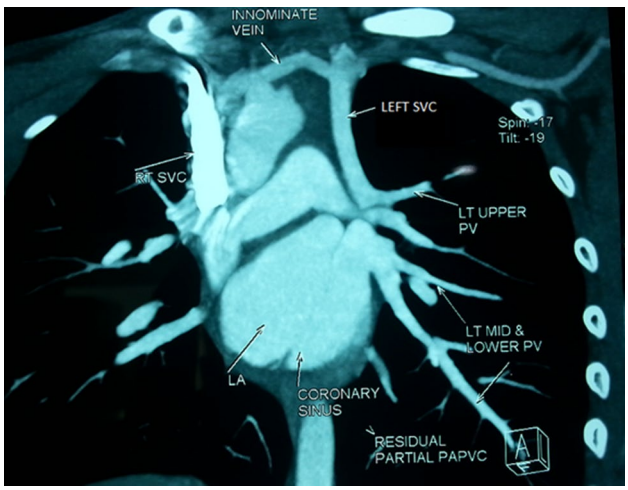

Fig. 2 Post-operative CECT images showing left upper pulmonary vein still draining into right atrium via left SVC, while the remaining pulmonary veins were draining into left atrium

air, no evidence of significantly increased pulmonary blood flow on post-operative echo and due to presence of a connecting vein segmental PAH was not a possibility, no surgical management was performed. He is still under follow-up and doing well.

Discussion

The commonest partial anomalous pulmonary venous connection (PAPVC) is right superior pulmonary vein attaching to the low SVC or the SVC-RA junction [1, 2]. In isolated right PAPVC, right pulmonary veins usually connect to derivatives of the right cardinal system: superior vena cava (SVC), inferior vena cava (IVC), or right atrium while in isolated left PAPVC, left-sided pulmonary veins usually connect anomalously to derivatives of the left cardinal system: coronary sinus and left innominate vein via connecting vein. This connecting vein has been called as a persistent left superior vena cava, but it usually does not make direct connection to the heart, so it cannot be identified by same name; a preferable term is anomalous vertical vein [3].

Bilateral PAPVC, that is a partial anomalous venous connection of both lungs, is very rare [1, 2]. The most common variant of bilateral PAPVC is probably that in which the left superior pulmonary vein attaches to the left innominate vein by way of an anomalous vertical vein, and the right superior pulmonary vein attaches to the SVC-RA junction and it is usually associated with intact atrial septum or with sinus venous defect [1, 2, 4]. The presence of bilateral PAPVC with left pulmonary vein draining into left SVC–>coronary sinus–> right atrium with the presence of ostium secundum atrial septal defect is as far as we know, not reported in the literature yet. There are several ways for the management of this complex malformation. For rerouting of right sided PAPVC, either baffling of the pulmonary vein into left atrium with augmentation of SVC or warden procedure can be performed. For left-sided PAPVC, different techniques have been reported, all involving an end-to-side or side-to-side anastomosis of the left upper pulmonary vein and the left atrial appendage [1, 5, 6]. The advantages of this procedure are: it does not require the institution of cardiopulmonary bypass and it can be performed through a left thoracotomy. However, it has a relatively high incidence of stenosis and was not suitable for this patient [7]. Other options are (1)ligation of left SVC near left innominate vein, cutback of coronary sinus and closure of coronary sinus opening with patch used for closure of ASD, (2) transection of left SVC above pulmonary vein opening and anastomosis of transected left SVC with left auricle in V–Y fashion to decrease the incidence of post-operative pulmonary venous obstruction, (3) performing left-sided bidirectional Glenn [6], and (4) correction of right PAPVC alone if left upper pulmonary vein is not draining major segment of lung. Various anatomical patterns and their management described in the literature are enlisted in Table 1. In our patient, left SVC was smaller in size as compared to right SVC, so it was ligated along with rerouting of right PAPVC into left atrium with pericardial patch, unroofing of coronary sinus into left atrium, and closure of atrial septal defect and coronary sinus opening was performed. On post-operative CECT scan, we found that left upper pulmonary vein was still draining into left SVC as we had ligated left SVC below it. To prevent this

Table 1 Various anatomical patterns and their management described in the literature

Sr. no.	Authors	Anatomical pattern described	Management described
1	Bexton RS [8] et al. (1984)	The right upper pulmonary vein (RUPV) draining into superior vena cava (SVC)/right atrial junction and left upper pulmonary vein (LUPV) draining into Left SVC	–
2	Ichihara T [9], et al. (1995)	The RUPV draining into SVC, and LUPV draining into the innominate vein with intact atrial septum	Warden procedure +_rerouting of RUPV + LUPV-left atrial anastomosis
3	Antonio F Corno [10] (1996)	The RUPV draining into SVC, and LUPV draining into the innominate vein with intact atrial septum	Baffling of RUPV to Left atrium + LUPV to LAA anastomosis
4	Kamota [11] T et al. (2007)	The RUPV draining into SVC, and LUPV draining into the innominate vein via vertical vein with intact atrial septum & pulmonary stenosis	Baffling of RUPV to Left atrium + LUPV to Left atrial appendage (LAA) anastomosis + Pulmonary commissurotomy
5	Sasikumar N [5] et al. (2014)	RUPV draining into right SVC-right atrium junction, LUPV draining into left SVC with sinus venosus atrial septal defect and a patent ductus arteriosus	Warden procedure + LUPV to LAA anastomosis + left bidirectional Glenn

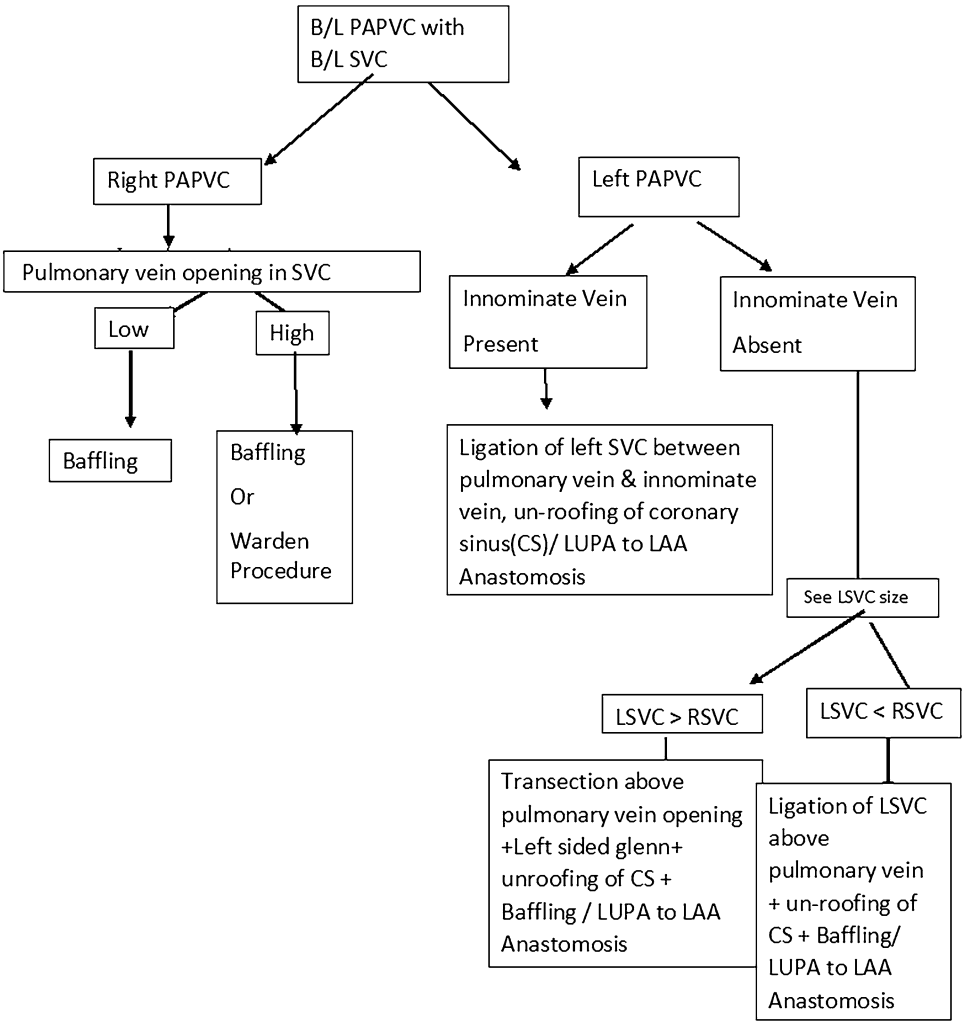

Fig. 3 Algorithm for management of bilateral PAPVC

complication, we recommend 1. preoperative CECT for these patients with accurate diagrammatic representation of all pulmonary veins and its branches and 2. intraoperatively, dissection of pulmonary veins up to the level of

azygous/hemiazygous vein draining into SVC or up to connecting vein should be done and the dissection should not be limited to the intrapericardial portion.

Conclusion

Bilateral PAPVC is very rare congenital cardiac anomaly in which surgical management needs to be individualized. The detailed anatomy must be delineated using echocardiography and/or CT angiography before deciding the surgical plan for successful correction of this rare cardiac malformation to prevent complication of residual PAPVC. The algorithm for surgical plan is described in Fig. 3.

Compliance with ethical standards

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Conflict of interest None declared.

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