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Anomalous Drainage of Persistent Left Superior Vena Cava to the Left Atrium: Case Series and Literature Review

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Abstract

Persistent left superior vena cava (PLSVC) draining into the coronary sinus is not uncommon, but to the left atrium is a rare condition. Such anomaly may be a cause of unexplained hypoxia in all age groups. It is often diagnosed accidentally during cardiovascular diagnostic work-up or in a context of other cardiac investigations. We report two cases of these rare PLSVC associations with subsequent medical and surgical management.

Keywords: Persistence left superior vena cava, Left atrium and management

1. Introduction

Amongst the anomalies of systemic thoracic veins, the persistent left superior vena cava (PLSVC) is the most common, with an incidence ranging from 0.3 to 2% in the general population [1]. The prevalence of PLSVC in individuals with congenital heart disease is estimated to be approximately 4.5% with variations amongst different studies [2,3]. Moreover, near to half of patients with PLSVC have other cardiac malformations, such as tetralogy of Fallot (TOF), atrial septal defect (ASD) or endocardial cushion defect [1, 2]. Conceivably, the drainage of PLSVC to the coronary sinus (right atrium), per se, is considered to be an innocuous variant of this pathology as it does not cause right to left shunt with its pathophysiological consequences. However, PLSVC draining into the left atrium (LA) is a serious condition as it causes blood mixing and

subsequent set of symptoms that necessitates surgical correction. Anatomically, there are three possible scenarios for the PLSVC to join the LA: through an unroofed coronary sinus, direct connection to the LA body, or through the left superior pulmonary vein [1]. Such anomaly is associated with an increased risk of cyanosis, intra cerebral abscess, heart failure, and embolic cerebrovascular stroke [1]. Moreover, when the PLSVC connects to the left upper corner of the LA body, it is usually associated with an absence of the partition between the coronary sinus and the LA a condition referred to as the “unroofed coronary sinus syndrome” [4]. This condition is particularly encountered in the setting of atrioventricular septal defects (AVSD), and Cor triatriatum [4]. Repair of all types of PLSVC is exclusively surgical. We report two cases of PLSVC associations with TOF and partial AVSD in the setting of bilateral SVC and small bridging veins.

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2. Case Reports

2.1. Case 1

A 6-year-old girl, 12.9 kg body weight, was diagnosed in early childhood to have non treated TOF; had a history of frequent cyanotic spells, finger clubbing (IV), oxygen saturation by oximetry was 60–70% on room air. Echocardiogram confirmed the diagnosis of TOF, bilateral SVC and vascular ring composed of right aortic arch, aberrant left subclavian artery. Since the coronary sinus couldn't be visualized, this raised the suspicion of PLSVC into the left atrium. To clear this cardiac catheterization was arranged. Diagnostic cardiac catheterization besides delineating further the anatomy, has confirmed PLSVC drainage to the LA (Fig. 1). Surgical repair was done via median sternotomy, in a contest of standard TOF repair [VSD closure, right ventricular muscle band (RVMB) resection and transannular patch] the PLSVC was isolated and identified. After coming off bypass, we encountered a saturation problem that could not be explained despite good surgical repair and near normal right ventricular pressures. We postulated that it is due to right to left shunt and mixing through the PLSVC. Therefore, a bubble contrasts transesophageal echocardiography (TEE) was done from the left-arm vein, which showed clear crossing of the bubble contrast to the LA confirming the drainage of PLSVC into the LA (Fig. 2a and b). This crossing disappeared after temporary ligation of the PLSVC.

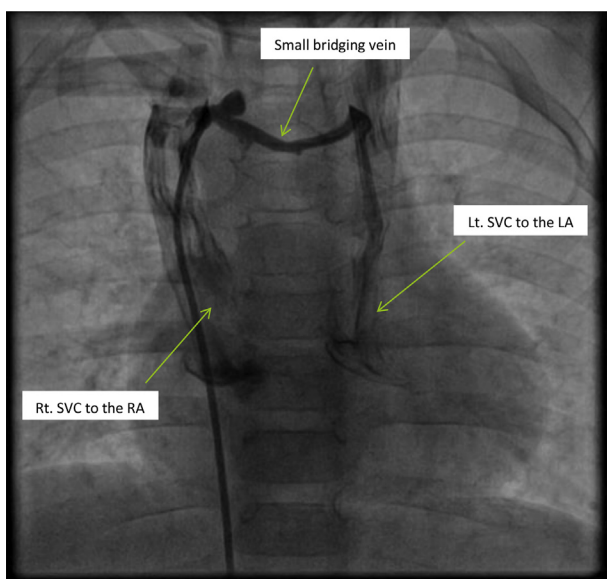


Fig. 1. Right SVC angiogram showed small bridging vein with good size left SVC draining into left atrium (Lt = left, Rt. = right, SVC = superior vena cava, LA = left atrium, RA = right atrium).

Baby's oxygen saturation has improved as well. The main concern was the size of the bridging vein and whether it can handle the venous drainage of the left side or not. Therefore, after a period of observation for signs of venous obstruction, the PLSVC was ligated permanently. The patient was transferred in a stable condition to the pediatric cardiac intensive-care unit (PCICU). She had a smooth postoperative course. Discharged home within 1 week, she is following-up in our pediatric cardiology clinic till last month and is doing well. Serial follow-up echocardiograms showed no residual cardiac lesion and patent left innominate vein with good flow to the right SVC. Oxygen saturation by pulse oximetry all the times was above 95% on room air.

2.2. Case 2

A five-month-old girl, the 5th child of healthy, non-consanguineous parents, at the age of 4 months, when her body weight was 6.9 kg, she was admitted to a local hospital due to fever and cough. Their echocardiography revealed transitional AVSD and mild pulmonary valve stenosis, so the patient was transferred to us. In our institution, the clinical examination was grossly normal apart from low oxygen saturation (85–90%), but no signs of respiratory distress. The chest X-ray depicted mild cardiomegaly and increased pulmonary vascular markings. Echocardiography demonstrated large primum atrial septal defect (ASD) almost common atrium, small restrictive inlet muscular VSD, small restrictive posterior muscular VSD, two separate atrioventricular valves, bilateral SVC and small bridging vein. Right SVC draining normally to the right side of the common atrium, coronary sinus was not dilated raising a possibility of anomalous drainage of left SVC. Bubble contrast echocardiography was done from left-arm confirming that LSVC draining into the left side of the common atrium just at the base of the left atrial appendage (Fig. 3). The case was accepted for surgical repair. Anomalous drainage of Left SVC was confirmed during surgery. It was a large vein connected to the base of the left-sided atrial appendage, with the small bridging vein connecting the right and left SVC. AVSD repair was performed and left SVC was divided at the level of the base of the left atrial appendage transecting a cuff of the left atrial appendage, which was anastomosed to right SVC. The innominate vein was augmented with an autologous pericardium patch. The repaired innominate vein looked like a native. The patient had a smooth postoperative course and discharged from the hospital after 5 days in stable

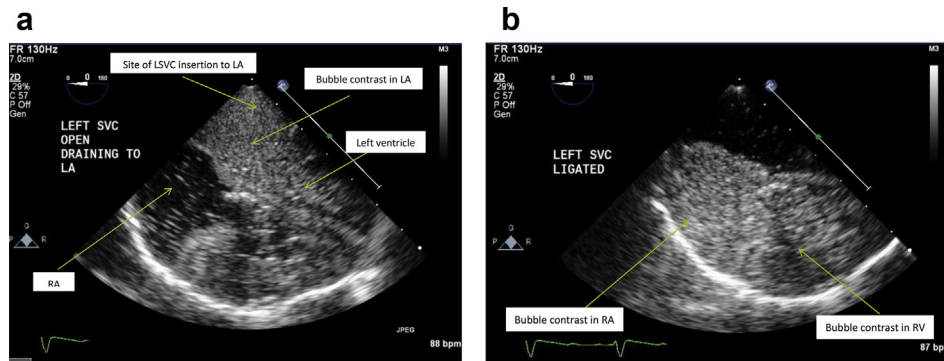


Fig. 2a. Intra-operative contrast bubble TEE study from left-arm showed left SVC draining to the left atrium just near the base of left atrial appendage before left SVC ligation. (LA = left atrium, RA = right atrium, LSVC = left superior vena cava). b. Contrast bubble intra-operative TEE study from left-arm showed normal drainage of the left innominate vein to the right SVC to the right atrium after left SVC ligation. (RA = right atrium, RV = right ventricle).

conditions. In an outpatient visit after three months from discharge the patient was hemodynamically stable her oxygen saturation on room air was above 95%. Echocardiography showed patent 'innominate vein' with good flow to right SVC, no residual ASD or VSD patch leak.

3. Discussion

The most common malformation of thoracic veins is the persistent left superior vena cava (PLSVC). It may drain into the coronary sinus in 3–10% of patients or to the left atrium in far less incidence in patients with congenital heart disease [2,6,7]. Usually, this anomaly does not cause hemodynamic effect unless there is a left to right shunt. In this instance, there might be a variable degree of systemic cyanosis, and increased risk of embolization and stroke [2]. Embryologically a left superior vena

cava arises when the left common cardinal vein and the left sinus horn merge to the right atrium. Failure of this merger leads to persistence of the connection between the left atrium and the left superior vena cava, which prevents the formation of the coronary sinus [2]. Since 1966, Mantini et al. have published a comprehensive review, classification and have proposed a surgical repair technique in a series of seven patients with coronary sinus anomalies, including its connection to the left atrium, pulmonary and systemic veins. He reported one patient PLSVC to the left atrium and unroofed coronary sinus with atresia of its mouth [6]. In 1986 Sand et al. described an alternative method of repair that consists of constructing a tunnel along the roof of the left atrium, thereby diverting the anomalous caval return into the right atrium [4]. In 1991 Wiles et al. reported two patients in whom a PLSVC drain to the left atrium with good size coronary sinus. One of these patients had tetralogy of Fallot (2 years old), while the other patient was nine months old with complete atrioventricular septal defect and pulmonary hypertension. Both patients underwent diagnostic cardiac catheterization, which confirm the diagnosis [2]. In 2008 Yousaf et al. described a PLSVC draining to the LA through the superior left pulmonary vein in a 71 years-old gentleman with a history of renal failure discovered during the placement of left internal jugular vein hemodialysis catheter [1]. In 2010 Ugaki et al. published his technique of repair of PLSVC by a division of left SVC from the left atrium and end to side anastomosis of PLSVC to the right atrium [5]. In 2012 Yesilkaya et al. reported a 69-year-old woman, who had PLSVC draining to the left atrium, absent right SVC, with bridging vein and normal heart anatomy, discovered accidentally during the multi-detector computed tomography [3].

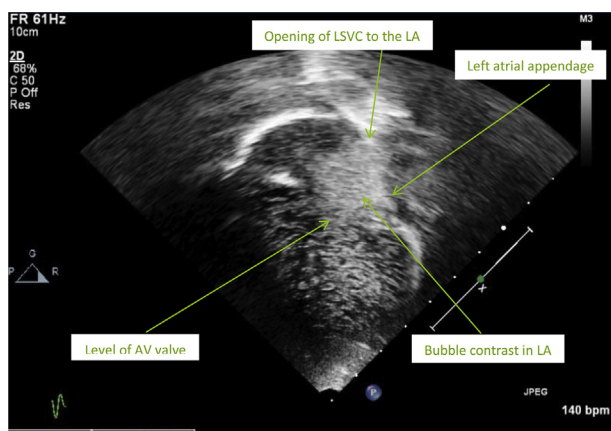


Fig. 3. Contrast bubble TTE study from left-arm showed left SVC abnormally draining into the left side of the common atrium at the base of the left atrial appendage. (LA = left atrium, LSVC = left superior vena cava, AV valve = atrioventricular valve).

In this study, we report our experience of the management of two patients with bilateral SVC, small bridging vein and truly PLSVC connected directly to the left atrium. The first patient had TOF and a vascular ring composed of right aortic arch and ligamentum arteriosum in the context of tetralogy of Fallot (TOF). After the echocardiogram, we decided to proceed with diagnostic cardiac catheterization to confirm the diagnosis. Then the surgical repair included vascular ring division and classic TOF full repair (VSD patch closure, RVMB resection and transannular patch). Initially, after coming off cardiopulmonary bypass we encountered a mild oxygen desaturation issue, which improved after PLSVC ligation. The post repair, TEE showed good surgical repair and the contrast bubble study after ligation of PLSVC showed the existence of the injected bubbles in the right atrium indicating adequate left side venous drainage, despite the smallish bridging vein (Figs. 2 and 3). In the second case the size of the bridging vein was rudimentary, so we applied the augmentation and re-implantation technique. Therefore, after the transitional AVSD repair, the PLSVC with part of left atrial appendage was resected and used as a patch to augment the small bridging vein. A small patch of autologous pericardium was used to align the new innominate vein. Both patients had a smooth course and discharged home in stable condition. Serial follow-up echocardiography showed patent left innominate vein with laminar flow to RA through the right SVC.

4. Conclusion

PLSVC draining to the LA is an extremely rare congenital cardiac anomaly that can cause low oxygen saturation in all age groups; this may have potentially serious complications as brain abscess, embolic cerebrovascular stroke due to right to left

shunt if not repaired. Most of the cases reported in literature are diagnosed accidentally during a routine investigation for other cardiac lesions.

The goal of this case series is to highlight the importance of recognizing this infrequent systemic venous anomaly and to stress on contrast bubble echocardiography from the left arm as a mandatory tool to confirm the diagnosis in cases of persistent left SVC with a normal size or absent coronary sinus. Once the presence of significant anomalous LSCV is confirmed, it must be rerouted or ligated.

Declaration

We have no conflict of interest to declare.

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